

DOCUMENT RESUME

ED 276 193

EC 190 959

TITLE Developmental Handicaps: Prevention and Treatment, II. A Cooperative Project between University Affiliated Facilities and State MCH/CC Programs.

INSTITUTION John F. Kennedy Inst., Baltimore, MD.; Johns Hopkins Univ., Baltimore, Md.

SPONS AGENCY Health Resources and Services Administration (DHHS/PHS), Rockville, MD. Office for Maternal and Child Health Services.

PUB DATE Aug 84

GRANT DHHS-PHS-MCH917

NOTE 135p.; For other documents in this series, see EC 190 958-961.

PUB TYPE Information Analyses (070) -- Reports - Descriptive (141)

EDRS PRICE MF01/PC06 Plus Postage.

DESCRIPTORS Adolescents; *Agency Cooperation; Clinical Diagnosis; Cooperative Planning; Cooperative Programs; Coordination; Delivery Systems; *Developmental Disabilities; *Diseases; *Health Services; *Interdisciplinary Approach; Needs Assessment; Pregnancy; Prenatal Influences; Prevention; Regional Programs; Universities

IDENTIFIERS Fetal Alcohol Syndrome; Iowa; *Title V State Maternal and Child Health Programs; *University Affiliated Facilities

ABSTRACT

Second in a series of three publications which address the prevention and treatment of developmental handicaps, the volume continues the effort to expand working relationships among university affiliated facilities (UAFs) and Title V Maternal and Child Health Programs. Five papers in Section I address developmental issues as of 1984: "Prenatal Diagnosis: The State of the Art" (L. Burney, A. Walker, K. Dumars); "The Fetal Alcohol Syndrome" (E. Ouellette); "Adolescents with Chronic Illness or Disability" (R. Nelson); "Methodology for Forecasting Service Needs for Chronically Ill and Developmentally Handicapped Persons" (P. Davidson and K. Roghmann); and "Regionalization of Health Service Systems" (A. Crocker). In Section II, E. Eklund updates linkages between UAFs and state and local Title V programs through a quantitative summary of reported collaborations as well as descriptive examples. A final paper by J. MacQueen ("The Integration of Public Services for Handicapped Children: Myth or Reality") outlines an interdisciplinary approach in Iowa to the full spectrum of assessment, rehabilitation, program planning, and integration of services. (JW)

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DEVELOPMENTAL HANDICAPS: PREVENTION AND TREATMENT II

A Cooperative Project
Between University Affiliated Facilities
and State MCH/CC Programs

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AAUAP

American Association of University Affiliated Programs
for Persons with Developmental Disabilities

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The UAF Network

University Affiliated Facilities (UAFs) are programs established to train personnel in the interdisciplinary approach to the delivery of services, to conduct research and to provide exemplary services related to persons with developmental disabilities and other multiply handicapping conditions. There are 51 programs in 37 states and the District of Columbia and many of the UAFs relate to programs in other states.

The American Association of University Affiliated Programs for Persons with Developmental Disabilities (AAUAP) is the national association of UAFs.

This project has been funded in part through Grant MCH917, Division of Maternal and Child Health, Bureau of Health Care Delivery and Assistance, Department of Health and Human Services through a contract between the AAUAP, the John F. Kennedy Institute, and Johns Hopkins University. The contents of this publication do not necessarily reflect the view or policies of DHHS.

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American Association of University Affiliated Programs

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Introduction

This is the second publication on approaches to planning for the prevention and treatment of developmental handicaps to be distributed by the national University Affiliated Facility (UAF) network with the support of the federal Division of Maternal and Child Health. It reflects a continuing effort to expand working relationships between UAFs and Title V Maternal and Child Health programs. The initial volume, published in 1983, met with a greater demand among a broader range of professionals than was originally anticipated; 5900 copies were ultimately distributed. Like its predecessor, the current offering is also divided into two sections, each with a specific focus.

The first section is comprised of five state-of-the-art papers, each a reasonably brief and easily readable up-to-date review of a topic of significance for Title V programs and their efforts in prevention, projection of service needs, and effective service programs for handicapped persons. Prevention, a continuing theme of this series, is specifically addressed in two of the papers. The first is a thoroughly current and succinct review of the rapidly developing field of prenatal diagnosis; the second deals with the fetal alcohol syndrome, the cause of a wide spectrum of developmental handicaps the full scope of which is only now being recognized.

The third paper deals with the needs of chronically handicapped persons in a particular age group, adolescence, a period demanding far more understanding and emphasis when handicap greatly amplifies the problems of an already difficult stage of development. It contains a plea for adequate needs assessments, approaches to which are reviewed in the fourth paper, on the methodology for projecting service needs for chronically ill and developmentally disabled individuals. The final paper in this section presents

several valuable examples of one approach to the coordination of resources in responding to perceived needs, namely regionalization of services.

The second section of the publication includes an update of the working arrangements, or linkages, between UAFs and Title V programs. This year's report is more extensive and, in addition to a quantitative summary of reported collaborations, includes excerpted material to convey the nature of the interactions, across a wide range of issues and from many parts of the country. Another attempt to move beyond mere numbers that is reflected here is the designation of what have been termed "new" or "emerging" relationships in order to move away from absolute comparison with more established relationships. In this section, too, is a report of the results of a survey on the scope and effectiveness of the use of state MCH and CC funded clinics as a clinical base for UAF training and on the reactions by those programs to the presence of UAF trainees.

Finally one potential approach to the goals of overall integration of programs for handicapped persons emphasized throughout this publication is presented in a paper by John MacQueen. It outlines an interdisciplinary approach in Iowa to the full spectrum of assessment, rehabilitation program planning and integration of services. Among many possible courses of action, it is one that merits careful scrutiny for ideas that could be adapted in other states.

In conclusion, we have tried to present to you what appears to be representative of the best trends in planning for the prevention and treatment of developmental handicaps, including ways in which the UAF network, state Title V programs, and other state agencies may interact to further these trends.

Chapel Hill, North Carolina
Silver Spring, Maryland
August, 1984

Harrie R. Chamberlin
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I.
DEVELOPMENTAL ISSUES:
THE STATE OF THE ART 1984

PRENATAL DIAGNOSIS: THE STATE OF THE ART

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Introduction

Prenatal diagnosis, or the ability to detect severe fetal anomaly in the midtrimester, is an achievement which has enabled thousands of couples at risk for malformation or genetic disease in their offspring to undertake pregnancies which otherwise would not have been attempted. The goal of prenatal diagnosis is to provide reassurance that a given problem does not appear to exist, or, in the few cases where a problem is found, to provide a couple the opportunity to review their options and to adopt the course of action which is most appropriate for them. When an abnormality is detected the options usually will include medical, educational, and psychological preparation for the birth of a handicapped child or, alternatively, interruption of the pregnancy so that another pregnancy or other reproductive options can be pursued.

In a few cases, a prenatal diagnosis has made it possible to intervene, either biochemically/pharmacologically, e.g., via alterations in maternal diet, or surgically, as with fetal urethral obstruction or hydrocephalus, in an attempt to ameliorate the fetal outcome. These efforts still are highly experimental and have had somewhat limited success. In most cases treatment is not yet possible. Fortunately, in 95% of cases, prenatal diagnosis provides reassuring information. A survey conducted by the Social Issues Committee of the American Society of Human Genetics (ASHG) revealed that the availability of amniocentesis encouraged families who were in a high-risk category to conceive, knowing that the pregnancy could be monitored. In fact, far more families in this situation delivered normal children than terminated

pregnancies for genetic reasons.

To date, myriad chromosomal disorders and well over 100 inborn errors of metabolism are identifiable by means of amniocentesis. In addition, numerous structural abnormalities have been identified through the use of ultrasound, various radiographic techniques, fetoscopy, and assay of maternal serum or amniotic fluid alphafetoprotein. Some resources that list disorders currently ascertainable are cited among the references, but techniques are advancing so rapidly that any published list is incomplete. For this reason, it is essential to contact a clinical genetics center whenever a question arises regarding the availability of prenatal diagnosis for a given condition.

This paper examines the process of prenatal diagnosis from the standpoint of genetic counseling considerations, indications for referral, and review of available technologies.

Genetic Counseling

Prenatal diagnosis should not be viewed as an isolated medical procedure. Because of the implications of discovering a fetal abnormality, the couple must be involved in a process of counseling that provides information regarding their potential risks for having a handicapped child, describes procedures which might be available for detecting these problems prenatally, and looks at the possible benefits, limitations, risks, and implications of any appropriate diagnostic techniques. The counseling process also provides an opportunity for the couple to explore their feelings about the diagnostic procedures and the possible outcomes. Following any studies, the counseling continues with a discussion and interpretation of results, review of options, and support for the couple. The obvious rationale for this counseling is to enable the couple to make an informed decision on how to proceed and to establish a support mechanism should an abnormality be found. In general, it

is much easier for individuals to comprehend and assimilate this vast amount of information prior to testing than when the couple is in a "crisis" situation immediately after a diagnosis is made.

Prior to counseling, it is imperative that the diagnosis/reason for referral be verified. Since many different disorders can look similar to the untrained eye and since family members are not always reliable sources of information regarding specific diagnoses, it is essential that every effort be made to verify the diagnosis with medical records and pregnancy and family histories. Misdiagnosis could lead to incorrect genetic counseling regarding recurrence risks and the availability of prenatal diagnosis, in turn creating false reassurance or unnecessary provocation of anxiety.

It has long been recommended that counseling be provided in a nondirective fashion so that the biases of the counselor, whether moral, religious, or ethnic, are not imposed on the counselee. Counseling should be provided so that the major emphasis is on the individual client's concerns. The benefits, limitations and risks of any appropriate procedures should be presented in an unbiased and truthful fashion, taking care not to omit any important details.

Preconceptual counseling obviously is of greater value than counseling after the birth of an affected child. Unfortunately, referrals rarely are made before a patient becomes pregnant. Referring physicians should be encouraged to obtain thorough histories from their patients before a pregnancy is initiated and to refer early so that the patients have the maximum number of reproductive options.

Whenever possible, both members of the couple should be counseled so that decisions regarding reproductive options can be made jointly. Since the issues that must be considered when making decisions frequently are quite complex and emotion-laden, it is optimal if both members can receive this information from the counselor first-hand so an opportunity for clarification

and emotional support will be available.

It is suggested that prenatal genetic counseling be provided by someone with specific expertise in this area. This individual may be a medical geneticist, a genetics associate/counselor, or a specially trained nurse, psychologist, social worker, or other professional. It may not be necessary to involve a medical geneticist or genetic counselor in routine cases such as those for advanced maternal age, provided that the physician or other professional who does the counseling is aware of the numerous issues which must be discussed and is cognizant of the appropriate manner in which genetic counseling should be provided.

When the genetic studies are complete, the couple must be able to discuss the results with someone qualified to explain the diagnosis, its implications and the available options. Since the couple will need some supportive counseling at this time, someone who previously has established rapport with the couple should be involved with the informing process. The genetics center can provide follow-up in terms of making needed referrals, confirming the diagnosis, and providing additional genetic and supportive counseling.

Indications for Prenatal Diagnosis

In the United States approximately 3-5% of all infants are born with a significant birth defect or genetic disorder. Unfortunately, the majority of these abnormalities are not detectable prenatally. Normal ultrasound studies and fetal chromosome analysis cannot guarantee a completely normal baby. Because of the limitations in what can be detected prenatally and the small risk associated with many diagnostic procedures, prenatal diagnosis should be reserved for situations in which the chance of abnormality and the value of the diagnostic information warrant the cost and risk of the procedure. Quantitatively it is fairly easy to calculate when a procedure's benefits outweigh

its risks; however, qualitatively this is not so simple. Some individuals may feel that the potential benefit of knowing early in pregnancy whether one is carrying a child with Down syndrome far outweighs the risk of miscarrying a pregnancy as a result of a diagnostic procedure. Others may feel that knowing whether or not the child they are carrying has Down syndrome is of no benefit when no in-utero treatment is available. It is not a physician's or a counselor's role to inform patients which procedures they must utilize; however, it is the professional's responsibility to inform them of the availability of these procedures when statistics show that the couple faces an increased risk of having a child with a type of anomaly that can be detected prenatally. Likewise, it is the physician's responsibility to refer a couple for genetic counseling even when they may be at increased risk of having a child with a problem not detectable prenatally.

Although prenatal diagnosis may not be available in every situation, a couple should be referred for a prenatal genetic evaluation when:

1. The mother will be 35 or older or the father will be 55 or older at the time of the child's birth.
2. Either parent has had a previous child with Down syndrome or another chromosomal abnormality.
3. There is a history of a relative with a proven potentially heritable chromosomal anomaly or mental retardation.
4. The couple has had a history of two or more miscarriages or infertility.
5. Either parent carries a balanced chromosome rearrangement (inversion or translocation).
6. There is a history of a congenital malformation in either parent or in a previous child.
7. There is a family history of a neural tube defect (e.g., spina bifida or anencephaly).
8. Both members of the couple are carriers for an autosomal recessive disorder (e.g., Tay Sachs, sickle cell, thalassemia, or PKU).
9. The mother is a known or possible carrier for an X-linked condition, e.g., Duchenne muscular dystrophy, hemophilia A, and X-linked mental

retardation with or without the fragile-X.

10. Either parent is affected with or has been shown to carry the gene for an autosomal dominant condition, e.g., achondroplasia, Marfan syndrome, tuberous sclerosis, neurofibromatosis.
11. The mother is an insulin-dependent diabetic, requires medication for epilepsy, or has a history of other potential teratogenic exposure (e.g., Rubella, X-ray, or certain drug use during pregnancy).
12. The parents are consanguineous (blood-related).
13. There is reason to suspect fetal abnormality on the basis of other studies (ultrasound, maternal serum alpha-fetoprotein, Rh titre).

Techniques of Prenatal Diagnosis

Fetal visualization

A variety of techniques have been developed to diagnose fetal structural malformations, anticipate pregnancy complications and assess fetal position and activity. These modalities may be used alone or in combination and also may be essential components of fetal sampling procedures.

Ultrasound. The use of high frequency pulse-echosonography for assessment of the fetus has largely replaced radiography in most obstetric situations. The technique has the advantage of being non-invasive and of being able to visualize both internal and external fetal structures. There is no maternal or fetal exposure to ionizing radiation and no apparent teratogenic or other risk (although long-term prospective studies still are in progress).

Two different types of scanners are routinely used for obstetrical ultrasound. The B-mode or "grey scale" equipment produces high resolution, static images, whereas real-time scanners enable visualization of fetal movement. Recently M-Mode scanners have been utilized for fetal echocardiography late in the second and in the third trimesters.

Ultrasound is useful both as a primary diagnostic tool and in conjunction with other diagnostic techniques, such as amniocentesis. It is used to verify

gestational age, locate the placenta, diagnose multiple gestation, assess fetal position and movement, identify gross fetal, uterine or adnexal abnormalities and to confirm a fetal demise.

Many fetal conditions associated with structural abnormalities have been successfully detected with ultrasound prior to 24 weeks. Among these are hydrocephalus and microcephaly (which may require serial studies), neural tube defects, renal agenesis or dysplasia, duodenal atresia, abdominal wall defects, cystic hygroma, fetal edema (which may occur with a chromosome anomaly) or hydrops, and limb defects. Fetal cardiac abnormalities have been ascertained in a few specialized centers during the midtrimester. Serial limb length measurements and assessment of fetal head/trunk ratio have made possible diagnosis of certain skeletal dysplasias. Identification of fetal abnormality in the third trimester may affect delivery management and can improve coordination of care for the malformed neonate.

It is imperative that ultrasound diagnosis, particularly of structural abnormalities, be attempted only by centers with specific experience in these types of mid-trimester diagnoses.

Fetoscopy. This technique is used in conjunction with expert ultrasound and is performed only by a limited number of skilled investigators in relatively few centers. Since it involves the transabdominal insertion of an endoscopic instrument considerably greater in diameter than the needle used in amniocentesis, the procedure entails greater maternal and fetal risk. Consequently the technique is appropriate only when the risk of the condition outweighs the procedural risk and when no other means of prenatal diagnosis exists.

Fetoscopy has been useful primarily as an aid to sampling fetal tissues such as blood, skin or liver for diagnosis of hemoglobinopathies, various white cell disorders, lethal ichthyoses or epidermolysis bullosa, and meta-

bolic errors in which the enzyme defect is not expressed in amniotic fluid cells. Fetal blood sampling via fetoscopy may be impossible when the placenta is anteriorly located. Dilution of fetal blood with amniotic fluid or contamination by maternal blood also may make accurate diagnosis impossible.

The usefulness of fetoscopy for anatomic diagnosis is limited by the very small field of vision ($2-4\text{ cm}^2$) provided by the endoscope. Ability to visualize the desired structures may be compromised by fetal position or movement, cloudy amniotic fluid, or veil of amnion obstructing the lens. Anatomic diagnoses have been accomplished in a number of disorders associated with limb defects and craniofacial abnormalities. Fetoscopy may provide a resolution when other data (e.g., ultrasound and AFP levels) conflict regarding the presence of malformation.

Radiographic visualization

1. Direct radiography. The utility of direct X-ray of the fetus for midtrimester diagnosis is limited by the degree of fetal skeletal ossification prior to 24 weeks, by difficulty in obtaining views of the fetus unobscured by the maternal skeleton and in some cases by inability to distinguish characteristic skeletal changes at this stage of fetal development. Most skeletal abnormalities which have been identified prenatally have been diagnosed fortuitously late in the third trimester during maternal pelvimetry performed for other indications. Attempts to diagnose skeletal dysplasias such as achondroplasia, asphyxiating thoracic dystrophy and thanatophoric dwarfism prior to 24 weeks by fetal radiographs have been largely unsuccessful both prospectively and retrospectively. Conditions associated with absence of long bones, such as Roberts syndrome or the thrombocytopenia, absent radius syndrome have been more amenable to diagnosis by second trimester X-ray.

Because the diagnostic capability of ultrasound in measurement of long bones and delineation of fetal structures has become increasingly sophisti-

cated, the use of this technique may be preferable to X-ray for most diagnoses, although fetal X-ray may be a useful additional tool to confirm and help interpret ultrasound findings.

2. Contrast radiography. Fetal radiographic diagnosis is more often accomplished with instillation of contrast medium into the uterine cavity. The process of injecting contrast material entails the same risks as inserting a needle for amniocentesis and thus should be attempted only by those with experience in this technique. Because additional complications such as fetal skin sloughs resulting from contact with contrast media and an increased incidence of fetal loss (0.5-3.0%) have been reported and because the long-term effects of fetal radiographic exposure* are not well documented, these procedures should be reserved for pregnancies known to be at high risk.

(a) Amniography utilizes a water soluble contrast material to opacify the amniotic fluid and outline the fetal surface. It has been successful in diagnosing meningomyeloceles not demonstrable by ultrasound following an elevated amniotic fluid AFP, anencephaly, omphalocele and gastroschisis, Meckel syndrome and a variety of other malformations. Since the fetus continually swallows amniotic fluid, a radiographic examination 24 to 48 hours following instillation of contrast medium may enable visualization of the fetal digestive tract. This technique has been useful in demonstrating obstructive phenomena and diaphragmatic hernia or agenesis.

(b) Fetography is possible only in the third trimester because the technique depends on absorption of an oily contrast material to the vernix caseosa. One day following instillation of the material, the fetal skin is well outlined, enabling visualization of external contours. Conditions accom-

*The levels of fetal exposure by diagnostic radiographic techniques usually are well under 1 rad and should be without teratogenic risk; however, the risk of childhood malignancy with 1 rad of intrauterine exposure has been estimated by some to be increased two-fold (which is an absolute risk of less than one percent).

panied by abdominal distention, such as polycystic kidneys, or by distortion of limbs, such as osteogenesis imperfecta, have been confirmed in late pregnancy using this technique. Complications which have been reported with the use of the iodinated contrast medium include minor effects on the fetal thyroid and respiratory distress resulting from fetal aspiration.

In general, fetal radiographic techniques are useful adjuncts to ultrasonography and amniocentesis and may provide information not obtainable via these other modalities. They should be used as confirmatory procedures prior to pregnancy termination whenever there is doubt as to the presence or nature of structural abnormalities suggested by other diagnostic studies.

Fetal Sampling

Amniocentesis. The technique of amniocentesis has been known since 1882, when it was first performed in Germany as a treatment for polyhydramnios. In the 1950s it became useful as a tool for monitoring fetal hemolytic disease in Rh incompatibility. In the 1960s it became possible to grow these cells in tissue culture, paving the way for the first prenatal diagnosis of a cytogenetic abnormality in 1967 and for the first diagnosis of an inborn error of metabolism in 1968. Since that time, the safety and efficacy of the technique have been documented by several major collaborative studies, and the ability to obtain amniotic fluid and fetal cells by this means has made possible the diagnosis of hundreds of different genetic conditions. While the relative safety of midtrimester diagnostic amniocentesis is well established, the technique requires greater skill than amniocentesis performed for therapeutic termination of pregnancy or for fetal diagnosis in the late trimester. For this reason, it should be attempted only by a physician specifically trained in midtrimester diagnostic amniocentesis who maintains his or her skill by performing a minimum of 100 procedures a year. The test is usually done between the 15th and 16th menstrual weeks following careful ultrasound exami-

nation. Prior to 15 weeks, there is a paucity of fetal cells and insufficient amniotic fluid for safe amniocentesis. Performing the procedure later than 20 weeks may not permit studies to be completed in time to allow the option of pregnancy termination. Usually a sample of 20-30 ml of amniotic fluid, obtained by means of a single, transabdominal needle insertion, is adequate for diagnosis. The most common indications for performing an amniocentesis are to rule out a fetal chromosome abnormality or neural tube defect. When a midtrimester amniocentesis is performed for other diagnostic purposes, arrangements should be made prior to obtaining the sample since the more specialized diagnostic procedures may be available in only a few centers.

1. Amniotic fluid studies. Analysis of various components of cell-free amniotic fluid is the basis for diagnosis of a wide variety of conditions.

(a) Elevated levels of AFP have been associated with open neural tube defects, fetal demise, congenital nephrosis, abdominal wall defects, twins, Turner syndrome, cystic hygroma, a variety of obstructive phenomena and numerous other congenital malformations. Low levels of AFP are normally present in amniotic fluid and decrease as pregnancy progresses. For this reason interpretation of the studies depends on accurate dating of the pregnancy and on normal standards which should be established by each laboratory.

(b) Assay of amniotic fluid cholinesterase (ChE) or acetyl cholinesterase (AChE) may be useful as an adjunct to AFP studies in diagnosis of an open neural tube defect.

(c) Diagnosis of numerous inborn errors of metabolism has been accomplished by assaying amniotic fluid for the deficient or abnormal enzyme. Pitfalls inherent in this technique include the possibility of spurious results due to admixture of maternal blood in the amniotic fluid or loss of activity of labile enzymes during transport of fluid. For this reason confirmation of the suspected diagnosis is usually achieved by studies on cultivated

amniotic fluid cells.

(d) Linkage studies through characterization of blood group substances in amniotic fluid have enabled diagnosis of a few disorders for which no gene abnormality has been delineated. Such studies rely on the close proximity of the gene for the condition to the gene determining a specific blood group characteristic (linkage). Prior to attempting diagnosis by this method, family studies must be conducted in order to establish the likelihood that an affected individual can be differentiated from one who is unaffected on the basis of blood group characteristics. Linkage between the genes for myotonic dystrophy and secretor status and between the ABO locus and that for nail-patella syndrome has been utilized for prenatal diagnosis of these conditions.

Linkage studies utilizing cellular components of the amniotic fluid also may provide a means of prenatally diagnosing conditions for which the actual gene defect has not been identified. Many HLA antigens are expressed on amniotic fluid cells, and tissue typing of these cells has enabled diagnosis of two conditions, 21-hydroxylase deficiency and C₄ deficiency, in which the gene is closely linked to the loci for HLA-A and -B. As with other prenatal diagnoses utilizing linkage, family studies must first be performed to establish tissue types associated with the abnormal phenotype. Fetal HLA typing has also been utilized for paternity testing. Because of the grave implications of misdiagnosis or of erroneous paternity determination, HLA studies of this nature should only be attempted by laboratories with experience in use of the technique for fetal diagnosis. (See Section 2[d] for a discussion of DNA polymorphism linkage.)

(e) Assay of testosterone levels in amniotic fluid has been used for fetal sex determination, although this method is considered unreliable. The technique may, however, be useful in resolving questions of laboratory error or maternal cell contamination of the sample. Conditions associated with

abnormally high testosterone levels, such as congenital adrenal hyperplasia, have been correctly diagnosed in this way (although linkage studies may now be more reliable).

(f) Optical density measurement of bile pigments in the amniotic fluid provides a tool for measuring the degree of hemolytic disease when fetal Rh incompatibility is demonstrated by rising maternal titres of anti-Rh₀ antibodies. Serial measurements are necessary to determine pregnancy management.

(g) Analysis of amniotic fluid phospholipids (L/S ratio) provides an important tool for assessing fetal lung maturity in cases of threatened premature delivery or maternal complications.

2. Amniotic fluid cells. Amniotic fluid contains a variety of cells derived from fetal skin, genito-urinary, respiratory and alimentary tracts as well as the amnion. The predominant cell upon culture is a fibroblast-like cell and its origin is still obscure. Presumably this fibroblast-like cell is derived from a "stem cell" which could be present in cells from any or all organ systems enumerated above. While diagnosis of chromosomal disorders can be accomplished using cultured cells of any cell type, the ability to diagnose an inborn error of metabolism is determined by the presence or absence of expression of the biochemical defect in the cell type studied. The observation of abnormal cell types in the amniotic fluid may indicate congenital malformation.

(a) Diagnosis of chromosomal abnormalities, such as Down syndrome and other conditions caused by an abnormal chromosome number, chromosomal rearrangements (translocations, inversions) resulting in deletion or duplication of chromosomal material, and certain conditions associated with chromosomal breakage (Bloom syndrome, ataxia telangiectasia, xeroderma pigmentosum or Fanconi pancytopenia) can be accomplished by cytogenetic analysis of cultured amniotic fluid cells. It may also be possible to identify some cases of

the "fragile-X" syndrome associated with X-linked mental retardation, but such attempts should be made only through cytogenetic laboratories with experience in the use of this technique for mid-trimester diagnosis. Although fetal chromosome analysis to rule out numerical or structural abnormalities is widely available, interpretation of the results is not always straightforward. It is important to confirm that the laboratory performing the studies will also be able to provide genetic consultation to the referring physician and/or patient in the event of an abnormal or ambiguous outcome.

(b) Fetal sex has been diagnosed by examining uncultured amniotic fluid cells for the presence of X or Y chromatin bodies. While this may provide a more rapid means of sex determination than cell culture and karyotyping, a benefit for example when the pregnancy is at risk for an X-linked disorder, the technique is impossible in a significant number of samples and has proven inaccurate in a small percentage of cases. For this reason, sex chromatin determinations on uncultured cells should not be used to ascertain fetal sex except as an adjunct in questions of maternal cell contaminations or possible lab error.

(c) It has been shown that activity of well over 100 enzymes associated with various inborn errors of metabolism can be demonstrated in cultured amniotic fluid cells. Prenatal diagnosis has actually been accomplished in at least half of these disorders and is potentially possible in the remainder. Most of these are rare autosomal or X-linked recessive conditions, and in most cases an at-risk couple is identified only after having had a previous affected child. It is not possible to diagnose disorders in which the underlying metabolic defect has not been characterized (e.g., cystic fibrosis, Duchenne muscular dystrophy) or conditions in which the enzyme is not normally expressed in amniotic fluid cells or is not active by 20 weeks gestation. Because diagnosis of a specific inborn error of metabolism may be available

only through one or two specialized laboratories and since preliminary studies may be necessary, it is essential that arrangements with the laboratory be initiated as soon as pregnancy is recognized.

(d) Prenatal diagnosis of a variety of conditions has now been achieved by direct analysis of fetal DNA extracted from amniotic fluid cells. If sufficient material is present in the amniotic fluid sample, culturing of cells is unnecessary and the time needed for diagnosis can be reduced. DNA studies were first used to demonstrate deletion of alpha globin genes in fetuses affected with thalassemia. More recently, DNA polymorphisms (variations in normal nucleotide sequence without changes in function) have been identified in DNA segments linked to the genes responsible for several different conditions. In many families a particular polymorphism is consistently associated with the mutant gene, making identification of affected individuals possible. Diagnosis of various hemoglobinopathies, e.g., sickle cell disease or B-thalassemia, can now be accomplished in most cases by DNA polymorphism studies, obviating the risk of fetal blood sampling. Identification of DNA polymorphisms associated with genes for conditions in which the enzyme defect cannot be demonstrated in amniotic fluid cells, e.g., PKU, or in which the underlying defect is not known, e.g., Huntington disease, has made their prenatal diagnosis potentially possible for the first time. As linkages between DNA polymorphisms and other genes are identified, this technique will become an increasingly important means of prenatal, presymptomatic, and carrier testing.

(e) Evaluation of cellular morphology and culture characteristics has been a useful additional tool in the diagnosis of a few specific birth defects. Macrophage-type cells which rapidly adhere to the culture vessel have been found in amniotic fluid when the fetus has an open neural tube defect. Similarly, rapidly adhering peritoneal cells may be observed when an

abdominal wall defect is present. This method should be used in combination with other, more proven modalities such as AFP and ultrasound.

Chorionic Villus Biopsy. The trophoblast is an extra-embryonic tissue mitotically derived from the zygote. It forms the outer wall of the blastocyst before implantation and is the source of the amnion, chorion and placenta. The villi on the nonimplantation side cease proliferation and by 14 to 15 weeks have disappeared. Between the 6th and 10th weeks of the pregnancy, however, there is still a fairly luxuriant fringe of villi surrounding the embryo. Since this tissue is derived from the original zygote, it accurately reflects the genetic constitution of the fetus and should provide a source of material for chromosomal and biochemical diagnosis. If this material can be obtained safely (and prospective collaborative studies to ascertain this currently are being organized) it may be possible to provide results of prenatal diagnosis of chromosomal conditions and some inborn errors of metabolism at 8-10 weeks following the last menstrual period rather than the 18-20 weeks now usual with amniocentesis.

At present, development of this technology is in its early stages. Preliminary reports indicate that a small sample of the chorionic villi can be obtained by directing a narrow catheter through the cervix to the site of implantation. It appears that this is best done utilizing direct ultrasound visualization to assure appropriate placement of the catheter. A tissue sample of about 5 mg is obtained by aspiration and the chorionic villi are isolated by microscopic dissection. The villi thus obtained can be used for direct chromosome preparations (since the tissue is rapidly dividing), for a source of DNA for diagnosis utilizing DNA polymorphisms (see above) or can be cultured for metabolic studies or more definitive chromosome analysis.

While the technique looks enormously promising, a number of questions still remain unanswered, and it may be a matter of years before the method is

widely available. The long-term effects of the procedure in terms of pregnancy complications or harm to the fetus are still unknown. Furthermore, it is not known what proportion of chromosomally or biochemically abnormal fetuses would spontaneously abort prior to 12 weeks, making such diagnostic procedures and therapeutic terminations unnecessary. It is unlikely that, even if proven safe, chorionic villus biopsy will completely replace current diagnostic modalities since the method provides no means of diagnosing malformations not associated with a chromosomal or biochemical error, such as neural tube defects or skeletal anomalies. However, the ability of chorionic villus biopsy to make diagnostic information available to at-risk couples nearly 10 weeks earlier than is presently possible may make prenatal diagnosis more widely acceptable.

Maternal blood studies: At present, all methods of fetal sampling (amniocentesis, fetoscopy and chorionic villus biopsy) involve intrauterine manipulation. Because this is not without danger, both to the mother and the fetus, these techniques are appropriate primarily when there is prior indication that there is increased risk for fetal abnormality. Unfortunately, most handicapped infants are born with no previous suggestion that the pregnancy was at risk. For this reason it is attractive to find non-invasive means of fetal assessment which are cost-effective in identifying those pregnancies in which more thorough diagnosis may be warranted. Since fetal cells and some fetal metabolites are known to enter the mother's blood stream, research has focused on ways that a maternal blood sample could provide a "window" to the fetus.

1. AFP, produced by the fetus, crosses the placenta and is present in low levels in maternal serum. Measurement of maternal serum alphafetoprotein (MS-AFP) thus provides a relatively non-invasive means of screening for neural tube defects and other fetal malformations. An abnormally high level of MS-

AFP will be present in about 5% of pregnancies tested at 16-18 weeks. In only a small proportion of these will a neural tube defect actually be present. Inaccurate gestational dates, fetal demise, twins and other types of fetal malformation may also cause an elevated MS-AFP. For this reason, additional studies, often including ultrasound and amniocentesis, must be performed. A specific protocol of procedures to be followed in the event of an elevated MS-AFP has been devised and must be adhered to in order to identify fetal abnormality correctly and to prevent the termination of normal pregnancies. MS-AFP screening should be performed only following informed consent and with access to appropriate counseling and diagnostic facilities.

2. Fetal red cells can be identified in maternal blood on the basis of size and of staining properties which depend on the presence of fetal hemoglobin. Such studies may be warranted if there is a question that isoimmunization of an Rh negative mother may have occurred, particularly as a result of amniocentesis.

Social and Ethical Considerations

It is likely that prenatal diagnostic techniques will continue to develop enabling detection of severe fetal anomaly earlier using less invasive methods. The emergence of a variety of new diagnostic capabilities in medical genetics raises numerous questions regarding the social and ethical implications of these technologies. It is crucial that further development of this technology be accompanied by thoughtful consideration of its appropriate use.

Hopefully, as technology evolves in this direction, our ability to intervene in more positive ways than pregnancy termination will also grow. Diagnostic acumen has already made possible identification of conditions which may be relatively mild or which are surgically correctable. It is likely that many more will be diagnosable in the future. This raises important ethical

questions concerning what constitutes an "acceptable" level of disability and to what degree prenatal diagnosis should be utilized as an avenue toward perfection rather than as a means of averting tragedy. Is it appropriate to utilize this ability to satisfy parents' desires for as perfect a child as possible? If we do so, what messages does this convey to handicapped individuals in our society? Public agencies should lead the way in examining these issues.

The advent of DNA polymorphism techniques is likely to greatly increase the numbers of disorders for which carrier detection, prenatal diagnosis and presymptomatic testing are available. However, some of these disorders may vary greatly in severity and while prenatal diagnosis of the condition may become possible, the use of DNA polymorphisms will not enable one to distinguish between a minimally and a severely affected individual. Furthermore, many of the disorders for which presymptomatic testing may become available cannot now be treated.

Although it may be beneficial in terms of reproductive planning for an individual to know that he will eventually develop a lethal, degenerative condition, the psychological impact of this knowledge may be devastating. Similarly, the benefits of carrier identification for genetic counseling may be outweighed by the risk of stigmatization and job or insurance discrimination.

Chorion villus sampling (CVS) must also be approached with caution. At present fewer than 800 of these procedures have been performed throughout the United States and less than half of these have been on continuing pregnancies. Only a small number of women have actually delivered, so no solid data are yet available on the risks or long-term effects of the procedure. The technique must still be viewed as experimental until questions regarding safety and accuracy can be resolved through well designed collaborative studies. Even

when this happens, it is unlikely that CVS will be widely available for some time and a means of prioritizing those desiring to avail themselves of the technique will have to be devised. Because CVS can diagnose fetal sex nearly 8-10 weeks earlier than is possible with amniocentesis -- at a time when the pregnancy may not be easily recognized -- the potential use of the technique for sex selection must be acknowledged. It is important to encourage public debate about whether this is an appropriate use of this procedure.

Implications for Title V Agencies and University Affiliated Facilities (UAFs)

Title V agencies and University Affiliated Facilities (UAFs), by virtue of their emphasis on maternal and child health and their concern about individual and collective health of society have a variety of responsibilities in the provision of preventive genetic services. Though Title V programs and the UAFs have differing mandates related to provision of service, education, and financial program support, the knowledge base in genetics required to carry out these varying mandates is quite similar.

Title V programs in their role as provider or fiscal agent for genetic programs must assume responsibility for monitoring the quality of services provided. As diagnostic procedures that were once viewed as experimental and appropriate for only a very select population, e.g., genetic amniocentesis, have become routine, and as public demand and utilization of these technologies has burgeoned, the delivery of these services has begun to change. Diagnostic tests once available only through genetics centers in tertiary care settings are increasingly provided by private individuals or commercial laboratories. This shift may be appropriate, since it may make genetic services more widely available and may allow tertiary centers to focus on development of improved techniques for diagnosis and management of genetic disorders. However, steps must be taken to ensure that quality care will be

preserved.

Three diagnostic areas appear to be particularly attractive to the private sector and each of them raises different quality control issues. The emergence of private laboratories which provide cytogenetic studies, both for pre- and postnatal diagnosis, has in some areas increased competition for referrals. Often, emphasis is placed on the rapidity of diagnosis, reduced cost, or convenience for the referring practitioner. Each of these are worthy goals, provided that they are not met by compromising care in the handling of specimens, by overloading existing laboratory personnel or hiring inadequately trained personnel, or by reducing the thoroughness or quality of the diagnostic procedures. Furthermore, cytogenetics laboratories should be recognized for what they are, and should acknowledge the limits of their expertise in other areas of genetic diagnosis and counseling.

A second phenomenon which bears watching is the increasing trend for the primary care physician, particularly the obstetrician, to want to provide the obstetrical component of prenatal diagnosis. At present this involves primarily amniocentesis, but with the advent of chorion villus biopsy, practitioners may wish to perform this procedure as well. With amniocentesis it has been demonstrated that risk to the patient is significantly increased when the physician has not had specific training in the technique or when it is performed relatively infrequently. Significant concerns arise around the relatively inexperienced practitioner who wishes to perform this procedure and about how the patient will be counseled regarding procedural risk. Since an increasing number of obstetricians also are performing ultrasound studies in their own offices, it will be important to monitor whether these procedures are being utilized primarily for obstetrical management or if a diagnostic capability beyond the limits of the equipment and experience of the practitioner is being implied.

A third area of concern is the advent of maternal serum screening for alphafetoprotein. The availability of commercial assays for AFP has opened the door for widespread screening and, while some states are attempting to establish protocols and regulate the use of this test, the potential for misuse and misinterpretation of results is great. Title V agencies must first educate themselves about the implications of MS-AFP screening and then take an active role in educating the professional community and public regarding the voluntary nature of the test and the importance of adherence to protocols.

Because Title V programs provide support for newborn screening programs, NICU follow-up, family planning and high risk obstetrical clinics, and clinics for a number of categorical diseases such as the muscular dystrophies, hemoglobinopathies or late-onset degenerative disorders, they are in an excellent position to identify families in need of genetic counseling and preventive services. It is important that, in these settings, increased emphasis be placed on obtaining patient and family histories which consider genetic factors that may place a couple at increased reproductive risk. In the evaluation of multiply handicapped children, particularly those with developmental delay and congenital malformations, the possibility of a genetic etiology must be considered and appropriate diagnostic studies arranged.

At present, as with many areas of health care, there is a maldistribution of resources for genetics services. Because many genetics units evolved within university medical centers they tend to be concentrated in metropolitan areas, and sparsely populated regions are often underserved. Title V agencies in less populous areas should be active in educating the private medical community regarding availability of preventive genetic services and the indications for referral. However, even geographic proximity does not ensure access, and many individuals living in areas that are well served do not receive services because of financial constraints or lack of knowledge of

their availability. It is incumbent upon Title V agencies to provide public education and to promote allocation of health care dollars to genetic services. While public health care programs must lead in the effort to contain medical costs, they must at the same time advocate the securing of realistic and optimal funding for all prevention services.

An overriding issue with the increased provision of genetic services by the private sector is the question of the adequacy of genetic counseling. Provision of such counseling is a technically complicated and time-consuming effort that does not reimburse well. There is great danger that this most essential component of prenatal diagnosis will be ignored or given short shrift. For patients to be asked to embark on a diagnostic process without fully understanding its implications or the magnitude of reproductive decisions they may be forced to confront is unethical and inhumane. It is crucial that public agencies be as aware of the adequacy of this aspect of privately provided genetic services as they are vigilant about the technical expertise of these providers.

The UAFs must be aware of the issues cited above and in addition, if the individual UAFs do not provide genetic services, including counseling, they should develop linkages with genetic programs to ensure service for their clients as well as training for their students and faculty. Lastly, Title V programs and UAFs should be active in examining the social and ethical implications of the new genetic diagnostic and therapeutic modalities, for public discussion is crucial to ensure that these technologies continue to have a positive influence on society.

It is apparent that these authors, along with others, regard genetic services as part of public health prevention programs. No other agency has been more involved and more concerned about the individual and collective health of society than Title V programs. It is therefore all the more crucial

that Title V and University Affiliated Facilities be knowledgeable regarding genetic services and participate in their further development.

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THE FETAL ALCOHOL SYNDROME

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Historical Review

Although the belief that parental alcohol intake may be harmful to the fetus dates back to antiquity, modern awareness of the ill effects of parental drunkenness on children began during the "gin epidemic" in 18th century England. The College of Physicians petitioned Parliament in 1726 for control of distillation, citing gin as "a cause of weak, feeble and distempered children." Further descriptions of early childhood death, feeble-mindedness and epilepsy in offspring of drunken parents appeared throughout the remainder of the century.

In 1899, Sullivan first demonstrated that children born to women alcoholics had a death rate 2.5 times that of children born to their female relatives. The main cause of death in this series was epilepsy (Warner & Rosett, 1975).

During the first half of the 20th century, there was little interest in this problem. Congenital malformations in children of alcoholic parents were first described in 1960. The pioneer work on the prenatal effects of alcohol was carried out by Lemoine and others in Marseilles in 1968 (Lemoine, Harousseau, Borteyro & Menur). They studied 127 offspring of alcoholic parents and described abnormalities which included low birth weight, short stature, delayed growth and development, cranio-facial and cardiac abnormalities, retarded intellectual and motor development, and impaired school function.

In the United States, Ulleland (1972) reported that 10 of 12 children born to 11 women alcoholics were small for gestational age. Development was

found to be retarded or borderline even when corrected for prematurity. Post-natal growth retardation also occurred. In 1973, Jones, Smith, Ulleland and Streissguth retrospectively examined eight unrelated children born to chronically alcoholic mothers and described the growth, morphological and developmental abnormalities for which they subsequently coined the term "Fetal Alcohol syndrome."

In a prospective study carried out at Boston City Hospital between 1974 and 1976, Ouellette and others found that the frequency of all abnormalities in offspring of heavy drinkers increased twofold as compared to abstinent and moderate drinkers. Thirty-two percent of infants born to heavy drinking women (at least 5 drinks on occasion and an average daily intake of 45 ml. per day of absolute alcohol) had some form of congenital malformation as compared to 9% born to abstinent mothers and 14% to moderate drinkers (more than 1 drink per month and less than the definition above for heavy drinking). Both major and minor anomalies were more common. Major anomalies rose from 3% and 2% in the abstinent and moderate groups to 17% in the heavy drinking group. Multiple congenital anomalies occurred in 3% and 5% of the abstinent and moderate drinking groups and rose to 20% among offspring born to heavy drinking women. Sixty-seven percent of heavy drinking women who significantly reduced their alcohol intake during pregnancy, or abstained altogether, had normal babies (Ouellette, Rosett, Rosman & Weiner, 1977).

Definitions

Fetal Alcohol Syndrome (FAS) refers to findings on offspring of women who chronically drink alcohol to excess during pregnancy (Jones et al. 1973; Jones & Smith, 1975; Ouellette et al. 1977). Minimum criteria for the diagnosis of FAS are:

- 1) Prenatal and/or postnatal growth retardation, with height, weight, and/or head circumference below the tenth percentile;

- 2) Central nervous system involvement, with neurologic abnormality, developmental delay or intellectual impairment;
- 3) Facial malformations, with at least two of the following three signs:
 - a) Microcephaly (small brain)
 - b) Microphthalmia (small eyes) and/or short palpebral fissures (the horizontal length of the eyes)
 - c) Poorly developed philtrum (the distance from the base of the nose to the upper lip) and/or flattening of the maxillary (cheek) area (Rosett, Weiner & Edelin, 1981).

The most common signs seen in FAS include microcephaly, short palpebral fissures, maxillary hypoplasia, narrow vermilion border of the upper lip, congenital heart disease of all types, renal abnormalities, spasticity, language and other developmental delays, poor feeding and failure to thrive. Children with FAS also frequently have attention deficit disorders, hyperactivity, easy distractibility, problems focusing, and a variety of learning disorders.

The full spectrum of clinical features currently recognized in FAS is shown in Table 1.

Fetal Alcohol Effects (FAE) refers to any abnormalities seen in children as a result of alcohol use by their mothers during pregnancy (Hanson, Streissguth & Smith, 1977). These include any of the physical features of FAS, as well as failure to thrive, attention deficit disorders, learning and developmental disabilities.

Scope of the Problem

There were 3,704,000 babies born in the United States in 1982. FAS is estimated to occur between 1 and 2 per thousand live births for the full constellation of features (3,700 to 7,400 babies), with the frequency of

partial expression (FAE) at possibly between 3 and 5 per thousand live births (11,100 to 18,500 babies). Of the known causes of mental retardation, FAS is believed to be the third most common cause of mental retardation and is about as common as Down syndrome (Smith, Jones & Hanson, 1976). FAS IS TOTALLY PREVENTABLE.

The full scope and gravity of chronic maternal alcohol abuse prior to and during pregnancy is still unknown. Effects produced range from mildly impaired to profoundly afflicted children, and some fatalities have been reported. The risk of producing an abnormal child for a mother with alcohol abuse is unknown. Some evidence exists that children are more at risk for abnormalities with each successive pregnancy in which the mother continues to drink heavily.

The majority of children show only some of the abnormalities described. FAS should be suspected when growth retardation and/or congenital anomalies are present in a child and a careful history of ethanol intake should be elicited from the mother. Unfortunately, there are no laboratory tests which are diagnostic of this disorder, nor is there any present capability of identifying abnormal fetuses by amniocentesis.

Animal Research. A large number of articles have been published on the effects of known amounts of ethanol fed to a variety of pregnant mammals. In general, findings have been consistent with clinical reports on humans. Fetuses and newborn animals born to pregnant females fed high doses of ethanol have weighed less than control animals (Martin, Martin, Sigman & Radow, 1977).

A number of studies have resulted in the production of fetal malformations which are strikingly similar to those seen in humans (Martin et al. 1977). In the most elegant of these studies, Randall fed an experimental group of mice a liquid diet with 25% of the total daily calories supplied by ethanol from gestation day five through gestation day ten (Randall, Taylor &

Walker, 1977). Isocaloric paired controls were utilized. A large number of skeletal, heart and limb anomalies were observed in fetuses of alcohol fed mothers. No such malformations were observed in control fetuses. Renal anomalies described in these experiments predated the recognition of these findings in human FAS cases. Hydronephrosis and/or hydroureter of varying degrees, both bilateral and unilateral were noted. Cranial anomalies were also found in the experimental group.

Subsequent studies have demonstrated that in animals the effects on offspring are dose related. The more ethanol given to the pregnant animal, the greater the effect on animal pups. Decreased fertility, increased pre- and perinatal mortality and increased fetal resorption have all been described in a variety of mammals whose mothers have been fed large doses of ethanol during pregnancy (Martin et al. 1977). Survival times of rats whose nursing mothers were fed ethanol has been diminished.

A number of studies have been carried out in which the behavior of laboratory animals whose mothers were fed ethanol during gestation has been studied (Krasiak, Ellis, Poschlova, & Masek, 1977; Shaywitz, Klopfer, & Gordon, 1976). Unfortunately, some of these studies have failed to control adequately for nutritional intake, making their conclusions suspect. Among their findings were reported delays in the appearance of developmental reflexes in the rat. Others have reported increased aggressive behavior in pups of alcohol fed mothers (Krasiak, et al. 1977). A syndrome resembling minimal brain dysfunction in humans has been reported in rat pups born to alcohol fed mothers (Shaywitz, et al. 1976). General activity was significantly greater in these pups early on, but by 26 days of postnatal life the experimental group was not different from controls.

Audiogenic seizures have been produced in resistant mouse strains following ethanol feeding of mothers during pregnancy. Seizures have often

been long lasting.

A small number of neuropathological studies have been carried out such as that of Baur-Moffett and Altman (1975). Results of these studies have shown that the body weight and brain tissue weight were reduced significantly in ethanol-exposed rats. In one study, the weight of the cerebellum was reduced nearly twice that observed for remaining brain tissue (Baur-Moffett & Altman, 1975). Nutrition was well controlled for in these studies.

Clearcut biochemical changes have also been demonstrated in fetal organs when alcohol was given to pregnant rats. Protein synthesis in fetal and neonatal rat livers was found to be inhibited in these animals. (Rawat, 1975, 1977). Maternal ethanol consumption also resulted in a decrease in hepatic total RNA content, RNA/DNA ratio and ribosomal protein content in fetal liver (Rawat, 1975). Fetal hepatic DNA content was not significantly affected. Prolonged maternal ethanol consumption by pregnant and lactating rats also resulted in significant changes in neurotransmitters in fetal brains (Rawat, 1977).

Brain myelin determinations have been carried out utilizing a rat experimental model. Ethanol fed pups consistently had smaller brain and body weight than controls. Ethanol fed prior to conception and throughout gestation resulted in a premature onset and slowdown of active myelinization. Myelin was chemically and morphologically immature heavy myelin.

Women and Alcohol

Drinking among American women has been rising steadily since World War II and now constitutes a major public health problem. A particular concern is that excessive drinking is rising at a rapid rate. The highest proportion of heavy drinkers among women is between the ages of 22 and 30, the age of peak reproductive years (National Institute for Alcohol Abuse and Alcoholism, 1975). The second highest period is the teenage years. Estimates of the

number of women alcoholics in the United States range between 1 million and 4.5 million (Keller and Gurioli, 1976).

Drinking During Pregnancy. Recent studies show that all women tend to decrease their alcohol intake during the early months of pregnancy, presumably from a combination of factors (Rosett, Ouellette & Weiner, 1976). The production of gastric distress by nausea and vomiting early in pregnancy has been cited as a cause of the decreased desire for alcoholic beverages at that time. Motivation for providing a maximum nurturing environment for the fetus may also be a contributing factor.

Pregnancy has been shown to be an excellent time to intervene with alcoholic women. Pregnant women have been shown to be responsive to counseling, and many alcoholic women have become abstinent for the remainder of their pregnancy when support and education have been provided (Rosett et al. 1976).

Maternal/Child Effects

In Utero. Alcohol passes rapidly from the maternal circulation to the fetus and assumes approximately the same concentration as in maternal blood (Dilts, 1970). Alcohol levels within the fetal circulation fall more slowly than in the maternal circulation, so that detectable levels of alcohol are still present in the fetus after the alcohol has been totally cleared from the maternal circulation (Wagner, Wagner & Guerrero, 1970). Not only does alcohol enter the fetal circulation, but it is also excreted into the amniotic fluid, where it remains in essentially the same concentration for several hours until slowly being cleared (Mann, Bhakthavatsalan, Liu & Makowski, 1975a, 1975b).

Perinatal Effects. Alcohol withdrawal syndrome has been reported in infants of chronic alcoholic women who were intoxicated at delivery (Nichols, 1967). Withdrawal symptoms have included jitteriness, coarse tremors of the

hands and feet, general restlessness, sleeplessness, excessive crying and hyperirritability. Unlike infants with narcotic withdrawal, gastrointestinal signs, such as vomiting or diarrhea, have not been observed.

Postnatal Effects. Ethanol and acetaldehyde levels have been measured in peripheral blood and milk of lactating women and animals. Ethanol has been found to reach human milk in a similar concentration to that in peripheral maternal blood, decreasing together with decreasing ethanol content of the blood, but acetaldehyde has not been detected in milk, even in the presence of considerable amounts in the blood (Kesaniemi, 1974).

Effects of Moderate Maternal Drinking

Although some studies have shown an increase in premature and other low birth weight babies with moderate drinking during pregnancy, several well-controlled prospective studies have failed to find an increase in the production of the clinical features of the Fetal Alcohol syndrome (Hanson et al. 1977). Safe levels of alcohol intake during pregnancy, if any, have yet to be determined. At the present time, it is generally recommended that pregnant women abstain from alcohol, although an occasional alcoholic beverage has not been found to be harmful to the fetus. Similar recommendations are made to nursing mothers.

Major Problems in FAS Children

Feeding Problems. In our experience, much of the postnatal growth retardation seen in FAS children is due to their active rejection of food. Video-tapes show that the children have increased but uncoordinated sucking and swallowing movements coupled with extrusion movements of the tongue, so that their intake of food is less than adequate and feeding times are prolonged. Even highly experienced foster mothers find these children extremely difficult to feed.

Generally, feeding problems are most significant during the first year of

life and gradually improve so that by the time FAS children are three or four years of age, they are consuming a more adequate diet. They continue to be highly selective in their food preferences.

Learning Problems. As the children grow older, developmental delays and mental retardation often become apparent. Children frequently have short attention spans and marked difficulty focusing on tasks. Learning disabilities and hyperactivity are sometimes the presenting problems in school age children.

Emotional Problems. Depression and other emotional problems frequently accompany these other problems. Mothers with drinking problems often provide inadequate nurturing and sometimes provide inadequate food, thus exacerbating problems of growth and development. Child abuse and/or neglect often contribute to poor learning and may result in the child's being removed from the home and placed in foster care. Marital problems are common. These secondary effects on FAS and FAE children may make it even more difficult for them to function adequately.

Recommendations for Early Identification and Treatment of FAS Children

Identification of Pregnant Women. Physicians and nurses in prenatal clinics, especially those dealing with adolescents, should routinely inquire about alcohol intake when women first seek prenatal care. Questions should be asked in a sensitive and nonjudgmental way. Although many individuals with alcohol problems will underestimate the amount of their drinking, a brief questionnaire developed by Rosett et al. (1981) has been found to be useful in identifying moderate and heavy drinkers in a nonthreatening manner.

TEN QUESTION DRINKING HISTORY FOR PRENATAL USE

Beer: How many times per week? _____

How many cans each time? _____

Ever drink more? _____

Wine: How many times per week? _____

How many glasses each time? _____

Ever drink more? _____

Liquor: How many times per week? _____

How many drinks each time? _____

Ever drink more? _____

Has your drinking changed in the last year?

Women who are discovered to be moderate or heavy drinkers should be provided with education and/or counseling concerning the risks of continued drinking during pregnancy. Women who continue to drink heavily during pregnancy are at high risk for delivering prematurely and should be considered for transfer to tertiary care facilities for delivery.

Early Identification of FAS Children. Many children with FAS are not being identified early; consequently, they are not receiving the special help which can assist them to receive adequate nutrition and to achieve their maximum developmental potential.

The staffs of neonatal nurseries, pediatric units and neighborhood health centers, pediatricians, family practitioners, Early Intervention Programs (EIPs), Head Start Programs and WIC nutritionists should all be familiar with common findings in FAS and FAE: congenital malformations, developmental delay, hyperactivity, poor feeding patterns and failure to thrive. Any of these signs may suggest the presence of FAS or FAE.

Parents of children with these problems should be asked about alcohol consumption during the pregnancy with the child. It is important to realize that an accurate history may not be obtained on initial questioning. Even women who drink moderately during pregnancy may feel guilty about it. As a relationship with the family builds, they may become more frank. An initial

denial of any alcohol intake during pregnancy should not discourage professionals from further inquiry. Questions about alcohol intake should be included in a list of several questions about the pregnancy, an approach which may also improve the reliability of answers.

Many children are first recognized as having FAS when they reach kindergarten or first grade and begin to show learning disabilities and/or inability to focus on their work. Elementary and special education teachers need training to suspect FAS and FAE as causes of these problems, particularly when the children have minor congenital anomalies.

Children with FAS and FAE usually require interdisciplinary evaluations. Treatment plans must include provision for help for the families if intervention with the children is to be successful. Social work and other personnel should become familiar with alcohol treatment programs within their areas and develop linkages with state programs for alcoholism in order to facilitate referrals of mothers to treatment centers.

Prepregnancy Education

If the problems of FAS and FAE are to be eliminated or significantly decreased, young women must become acquainted with the risks of moderate and heavy drinking during pregnancy prior to the time they become pregnant. Recent animal research raises the possibility that, in the heavy drinker, at considerable risk for craniofacial abnormalities and significant retardation in her offspring, the most serious embryonic damage may occur at the time of gastrulation, three weeks following conception in the human being and thus often before the pregnancy has been clearly recognized (Sulik, 1984). Therefore, although it is important to provide education and counseling during pregnancy, preconception counseling is not only preferable but, in some cases, may be essential. The FAS Prevention Program at the Shriver Center UAF has

developed educational materials which are presently being included in junior high and high school health programs. Similar programs should be developed for young women in college and in the work force.

Staff in birth control clinics and adolescent clinics can play an important role in educating young women about FAS. Young women who are sexually active and who do not use birth control should limit their alcohol intake. Women's support groups may also play a useful role in education about FAS and FAE.

Public education about FAS can be enhanced by distributing brochures and by placing public service announcements on radio and television. Unfortunately, however, many of the brochures and public service announcements now in use do not stress the need for preconception counseling, particularly for heavy drinkers. They, instead, tend to convey the general message that, if one is pregnant and has an alcohol problem, one should seek help. As already mentioned, there is now suggestive evidence that, when alcohol intake is high, severe damage may occur before a woman is aware that she is pregnant. This caution should be included in public education materials.

Implications for Title V Programs and University Affiliated Facilities (UAFS)

Adolescent Pregnancy Programs. The inclusion of educational programs about FAS and FAE in adolescent pregnancy programs is critical. It is important to make teenagers aware that if they have been drinking heavily early in the pregnancy, there is a good chance that they can have a normal baby if they stop drinking as late as the third trimester (Rosett et al. 1981). The risk of prematurity and low birth weight babies appears to be correlated with drinking habits late in pregnancy, whereas congenital malformations, which clearly occur during the first trimester, have thus far been much more likely to occur in offspring of chronic alcoholic women. Teenagers, who are less

likely to have been abusing alcohol for many years, thus are also less likely to have babies with morphological abnormalities. They greatly decrease their risk of having premature and/or low birth weight babies if they abstain from alcohol during the second half of their pregnancy.

Early Intervention Programs. Staff in EIPs are in a unique position to identify young children with FAS and FAE on the basis of their physical features and to suspect it in children with otherwise unidentified developmental delays. They are also then able to provide treatment for children and support for families as part of their ongoing programs.

Women, Infants and Children (WIC) Programs. WIC nutritionists are sometimes the first professionals to identify children with failure to thrive (FTT) and other nutritional problems. FAS should be considered as a possible cause of the FTT. They must develop expertise in eliciting alcohol histories from mothers of children with FTT and learn to refer children and families with suspected FAS and FAE for appropriate evaluation and treatment.

Maternal and Infant Care (MIC) Programs. Professionals in these programs can provide parent education about excessive drinking during pregnancy, as well as identify cases of FAS and FAE.

Role of University Affiliated Facilities. UAFs can serve as regional resources for the diagnosis and treatment of FAS and FAE by providing assessments of these children by interdisciplinary evaluation teams and specialized services, such as feeding clinics. They can develop lists of regional resources of alcohol treatment centers for women and make these available to professionals in all fields who may encounter women who need help.

They can utilize their expertise in training to develop teaching packages for professionals likely to encounter children with FAS and FAE and to provide workshops for personnel in newborn nurseries, pediatric clinics, EIPs and Head Start Programs, and for WIC nutritionists, elementary and special education

teachers, and learning disabilities specialists.

Surprisingly, many alcohol counselors know little about FAS and FAE. UAFs can provide them with information about these disorders and train them to educate the women they serve.

UAFs can also develop curriculum materials for use in junior high and high school health programs. They can prepare brochures and other instructional materials about FAS for use in adolescent, birth control, and prenatal clinics. They should develop linkages with state departments of prevention, alcohol treatment centers, and state legislators to increase their awareness of the frequency of FAS and FAE as a cause of long-standing disabilities. They can have a direct impact on the development of education and prevention programs on a state and regional basis.

Summary

FAS and FAE are common and totally preventable causes of mental retardation and developmental disabilities. Their manifestations range from a recognizable cluster of birth defects to more subtle signs, such as problems with attention and learning. UAFs can play an important role in educating professionals likely to encounter children with FAS and FAE to recognize these conditions. They can also serve as regional resource centers for the early identification and treatment of FAS children.

TABLE 1
CLINICAL FEATURES OF FAS
TABLE OF ABNORMALITIES

GROWTH ABNORMALITIES

Prematurity
Intrauterine Growth Retardation
Postnatal Growth Retardation
Short Stature
Diminished Weight

CRANIOFACIAL ANOMALIES

Microcephaly
Eye Abnormalities
Epicanthal Folds
Telecanthus
Short Palpebral Fissures
Corneal Opacity
Ptosis
High Myopia
Strabismus
Tortuosity of Retinal Vessels
Flattened Nasal Bridge
Abnormally Formed Ears
Maxillary Hypoplasia
Narrow Vermilion Border of Upper Lip
Small Mandible
Cleft Palate

JOINTS AND LIMB MALFORMATIONS

Limitation of Elbow Extension
Phalangeal Anomalies
Small Nails
Clinodactyly
Abnormal Palmar Creases
Dislocated Hips

CARDIAC ABNORMALITIES

Atrial Septal Defects
Ventricular Septal Defects
Tetralogy of Fallot
Patent Ductus Arteriosus
Aortic Arch Interruption Type A
Peripheral Pulmonic Stenosis

RENAL ANOMALIES

Hydronephrosis
Single Kidney
Hypoplastic Kidneys

FUNCTIONAL ABNORMALITIES

Neonatal

Poor Suck
Tremulousness

Postnatal

Developmental Delay
Mental Retardation
Poor Gross Motor
Coordination
Learning Disabilities
Hyperactivity
Decreased Attention Span

OTHER FINDINGS

Hydrocephalus
Neural Tube Defects
Single Umbilical Artery
Noonan Syndrome
Klippel-Feil Anomaly
Capillary Hemangioma
Abnormal External Genitalia
Accessory Nipples
Adrenal Cortical Carcinoma
Spastic Diplegia

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ADOLESCENTS WITH CHRONIC ILLNESS OR DISABILITY

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The Phenomenon of Chronic Conditions in Adolescence

Medical research provides increasing insight into the health of the very young and the very old. The beginning and ending of life are hallmarks in the advance of medical intervention. Infant mortality falls; life expectancy rises.

The intervening years between birth and death are the obvious focus of much of health care. In less than three generations a host of killing and disabling diseases have been largely vanquished in American society - dysentery, polio, and tuberculosis are common illustrations. Acute and chronic conditions have become amenable to prevention, amelioration or cure. In selected circumstances the impact of treatment is dramatic, as in the care of very low-birth weight premature infants or individuals with end-stage renal disease. Other thousands of persons benefit from therapy which, while not curing the disease, makes functional daily activity possible.

The variety of chronic health conditions that limit function or well-being is almost without limit. Their impact can be particularly devastating during the time when the human organism is undergoing the major transition from childhood to adulthood (Wehlage, 1983). Youth with chronic illness or disability confront an even more uncertain change. The arduous adolescent process of self-identification, family separation, career preparation and adult relationship-building may be compromised. Future options may be dependent on the status of the health condition rather than the ability or initiative of the youth. Dependency is maintained on health professionals and

institutions. Financial hardship related to the costs of continuing care may plague the family of the youth. An uncertain medical prognosis may undermine emotional health.

There is a justification in contemplating youth with chronic illness or disability as persons adrift. The protective mantle and bond of the parent-child dyad are disrupted, yet the freedom and security of adult life appear unattainable. The youth cannot be "at home" in either locus. Health care reimbursement will illustrate this dilemma. As the child of an employed parent, a typical youth will be eligible for insurance coverage as a dependent. This coverage will extend into the young adult years, to age 21 or beyond, as long as the offspring remains dependent on the parent. For youth who have some chronic diseases the continuity of health insurance is a necessity. A parent may have foregone a new career opportunity to assure continuation of health insurance for the child. When the youth faces a decision to seek employment and economic independence the availability of health insurance may be a critical factor. Many entry-level jobs either do not provide comprehensive insurance or only offer contracts that exclude pre-existing medical conditions. Fiscal independence from family may be possible only if the individual is eligible for assistance from a tax-supported program. The youth may then experience a transfer of dependence, from parent to government.

The facilitation of health for youth with chronic illness or disability may be the great, if not last, frontier for concerned parents and professionals. It is imperative that there be an extension of the resources invested in the well-being of young children to include adolescents struggling with the very same conditions and with their personal identities.

A poignant example of this adolescent dilemma is the situation of young people with Prader-Willi syndrome, a developmental disorder typified by obesity, cognitive handicap, hypotonia, and hypogonadism. This syndrome, first

described in 1956, has attracted considerable attention due to the compulsive eating habits of affected individuals. The successful prevention of morbid obesity has only been possible through careful dietary supervision during childhood. Furthermore the behaviorable temperament of the children, characterized by stubbornness and a predilection to tantrums, is a concern to most families.

A clinical experience with young people with Prader-Willi syndrome suggests that adolescence is a particularly difficult time. Many of the youth have the intellectual ability and motivation to live apart from their families as they approach adulthood. These youth can generally manage their own self care needs and use public transportation in going to and from school or work. Self-control of inappropriate eating patterns, however, remains elusive for most individuals. Most intervention programs have emphasized the external control of dietary intake rather than personal motivation to use food in moderation. Food remains a preoccupation and often establishes the basis for social interaction, a frequent deleterious situation if the individual engages in food-stealing or other inappropriate behaviors.

Youth with this syndrome are often denied the opportunity to maximize their independent living skills due to this problem. A more sensitive network of community-based services may encourage greater independence.

Social Policy and Adolescents with Chronic Health Conditions

Health care of adolescents with chronic conditions has emerged as a focus of social policy. Between 1970 and 1976 legislation relevant to the health of handicapped adolescents was enacted in 45 countries (Paxman, 1982). These laws center on prevention, detection and reporting, treatment and rehabilitation. Four distinct federal legislative initiatives in the United States form the basis of current American law.

The Rehabilitation Act of 1973 (Public Law 93-391) prohibits discrimination against qualified handicapped people in programs, services and benefits that are federally funded. Health professionals and facilities accepting payment for services from federal programs are included in these provisions, as stated in Section 504 of the Act. The legislation is widely accepted as the primary civil rights legislation for handicapped persons in this country. During 1983, major controversy emerged when the federal administration promulgated regulations, based on Section 504, prohibiting the denial of medical care and nutritional support to handicapped newborn infants. The extension of that principle to older children and youth has not been explicitly interpreted by the courts, but is implicit in analogous health care situations for adolescents.

The Developmental Disabilities Assistance Act of 1975 (Public Law 94-103, amended in 1978 by 95-602) established protection and advocacy systems and representative councils for developmentally disabled people in each state. Furthermore, the Act made a contribution to societal definition of disability by emphasizing the functional issues of a developmental disorder rather than a medical diagnosis. A sizable number of individuals with developmental disorders have concomitant physical conditions, including epilepsy, nutritional abnormalities, and chronic bowel and bladder deficiencies. Vast changes are occurring in the planning of community services for persons with developmental disabilities which are relevant to health care providers and facilities.

The Education for All Handicapped Children Act of 1975 (Public Law 94-142) assures all children and youth in the United States an appropriate and free education from 5-18 years of age, and state law may extend this on either end of the range. A major consequence of this legislation is the necessity for schools to continue to serve youth who might in earlier years have left school discouraged by the lack of appropriate programs, thus compounding

maladjustment to adult life.

Finally, the Maternal and Child Health Services Block Grant of 1981 (Public Law 97-35, a part of the Omnibus Budget Reconciliation Act) provides funds to states for a variety of purposes, including necessary corrective, surgical, and rehabilitative services for children and youth who have handicaps or health conditions which might lead to a handicap. Most of these funds are administered by Crippled Children's Services programs, agencies having considerable variability among the states. In almost all states, however, these programs have expanded eligibility and services beyond early emphasis on musculoskeletal disorders. Therefore a legislative framework exists to approach the needs of adolescents with chronic conditions (DeLong and Lifchez, 1983).

The Scope of Chronic Health Conditions Among Adolescents

National Health Interview Survey data suggest that 4.7% of all youth ages 12-17 years are limited in activity by a chronic condition (Kovar, 1982). In the entire American population that percentage represents approximately 1,176,000 adolescents. Days of restricted activity, confinement to bed and school lost were highest in families of low income and less parental education. Across all age groupings, children from lower income families had a greater number of lost school days. Despite these findings, data from the same survey indicated that adolescents had the fewest contacts with physicians per year of any other age grouping of children. A reasonable conclusion might be that youth utilize required health care services less than younger children.

Estimates of the prevalence of chronic diseases among youth vary widely in the literature due to several factors, including changes in the natural history of diseases, variability in definition of medical conditions, altered

epidemiological sampling techniques, true variations in incidence, and differing birth rates. A preponderance of studies concentrate on the prevalence of chronic conditions at birth, in early infancy, or middle childhood. Reports with age-specific prevalence rates are scarce. When survival achieves dramatic improvement, as in chronic renal disease, there may be major public interest (Adams, 1978). Recent mortality figures (Vital Statistics of the United States, 1979) shockingly report that the leading causes of death among adolescents and young adults are not the physical diseases, such as cancer, heart disease, influenza or diabetes. The three top ranking causes of death are accidents, homicide, and suicide. Together they account for four times as many deaths in this age group as the next seven causes of death (67,550 versus 16,700).

These mortality statistics suggest an urgent opportunity for the review of social policy. They share a common attribute: the potential for prevention. Accidents among adolescents most commonly result from motor vehicle injury or poor judgment in recreational activities. The use of alcohol, the understanding of safety issues, and lack of adult monitoring are all contributing factors. Homicide frequently occurs in a context of family violence or is associated with other less serious criminal behavior. The widespread availability of handguns and other weapons increases the risk of death in situations where less serious injury might otherwise occur. Youth committing such crimes often were not previously recognized as having emotional or behavioral problems. Inadequate recognition also contributes to the eventual success of suicide attempts in this age group. A societal solution to any or all of these causes of mortality must involve enhancement of family and adolescent education and an extension of behavioral diagnosis and support services. The death of a young person in a family generally leaves a permanent scar on parents and siblings that extends the impact of the death into the

lives of innumerable persons.

In contrast, children who are born with or acquire chronic handicapping conditions are experiencing increased life expectancies. While the incidence rates of individual diseases or syndromes may be low, the cumulative impact of these chronic conditions on families and society is immense. That impact is the result of the need for frequent and continuous special services through childhood into adult life. The development of an effective social policy has been hampered by the fragmentation of effort among families, service providers, and advocates who are concerned with individuals with chronic disorders. Most advocacy has centered on disease-specific programs.

As mentioned, limited information is available regarding the prevalence of the various chronic diseases in adolescence. Surveillance of selected congenital malformations by the Center for Disease Control suggests that since 1970 the incidence of certain major birth defects is slightly increased (MMWR, 1979). The rate of meningomyelocele decreased from 7.55 to 4.97 cases per 10,000 total births, an annual decline of greater than 6%. Small decreases were reported for congenital anomalies of the gastrointestinal tract and Down syndrome. Slight increases were noted for certain congenital heart defects (especially those associated with prematurity), hip dislocation and renal agenesis. Such data may suggest a small diminution in individual youth with selected birth defects during the next generation. That trend, however, is likely to be countered by the improved survival resulting from advances in medical treatment and society's general commitment to expend relatively huge sums of money to maintain the life of persons with catastrophic conditions.

Public Health Programs

To this time there has been little emphasis in public health expenditures on the adolescent population. A review of federal health services grants in

1980 (Zwick, 1981) identified only a single categorical grant for this population, targeted on adolescent pregnancy services. This community health services program commanded only \$6.5 million, less than 0.5 percent of the \$1.34 billion in federal expenditures that year. There were no specific programs relating to adolescents with chronic illness or disability, although this population was probably the beneficiary of other maternal and child health, mental health, and developmental disabilities projects.

Most public health program services for children and youth with chronic illness have depended upon eligibility by diagnosis and financial need. Beginning in 1935 with the passage of Title V of the Social Security Act, the Federal government, acting through the states, undertook the assurance of more comprehensive and accessible services to children and youth with chronic disease. Much of this effort initially centered on corrective or therapeutic treatment of children with physical disability. These children had the "fixed deformity" characteristic of musculoskeletal disorders and birth defects. State Crippled Children's Services (CCS) programs were organized to identify such children and assure access to service. However, due to limitations of direct service efforts in public health, the exclusive orientation to poor families, and persistently small appropriations, most state CCS programs focused on only a segment of the potentially eligible children. With time the scope of the programs included further medical diagnoses as efficacious treatment became available. An illustration of the expansion of these programs was the inclusion of congenital heart disease into most of them in the 1950s, following the introduction of successful open heart procedures and the advent of heart-lung bypass. Chronic illnesses such as cystic fibrosis and hemophilia also have been included as therapeutic approaches to these disorders evolved. Despite such changes the programs in most cases have maximal penetration into target populations of only 30-40%, meaning that the majority of

children and youth with chronic illness do not benefit from these efforts (Ma, 1979). It has been argued that not all youth with chronic illness require public health services since many families have the resources to obtain these services independently. In fact the denominator of need for specialized health care is rarely understood. This is due in part to our lack of knowing the true financial burden imposed by the costs of chronic illness on many families. It is further hampered by our primary lack of epidemiological information on the prevalence and severity of many of these illnesses.

Regardless of the specific pathology of the condition, most chronic illnesses share a basis of similarity relevant to determination of social policy. Almost by definition chronic illness requires costly extended care that is provided by specialists in medical centers. There is a daily burden of care imposed upon families that is accompanied by some discomfort or even pain for youth. Frequently these illnesses are progressive or at least are not curable, creating a sense of uncertainty as families plan their futures. Because of the low incidence of most of the conditions there is a poor understanding by the public and families themselves of the nature of the illness.

The health care of chronically ill youth therefore necessitates a careful review of the characteristics of current care and services. Whenever that care lacks coordination, comprehensiveness, or continuity, there is a significant risk that not only will the care be less than maximally effective, but also that secondary complications of the chronic illness will jeopardize a family. In view of the current public temperament regarding the consumption of resources in the health care system in general and the ineffectiveness of many public agencies in problem-solving, it is somewhat dubious that a major new effort to reorganize the service system will be launched in the foreseeable future (Peoples and Miller, 1983).

The options available presently center on the transfer of resources

within the system to provide better care. In order to assure greater access and comprehensiveness, such a transfer inevitably means the shift of dollars from high-cost services (hospitalization or residential care); such a transfer is not readily accomplished. The introduction of new technologies motivates institutions caring for youth to maintain their operational functions and maximize their investment in facilities and staff. Furthermore the reimbursement system has generally discouraged the use of ambulatory or noninstitutional services although it can be demonstrated that adoption of such services as an alternative to residential care is much less expensive in the long term. Redirection of providers, especially physicians and other decision-makers, to alternative models is critical in the shaping of a new system. Similarly, detailed studies must show that alternative financing of outpatient service will be critical and convince third party payers, both public agencies and private health insurers, that such models are not only desirable and necessary, but also might lessen the spiral of escalating health care costs for this population of youth. Even substantially innovative financing models, such as prepaid health insurance plans targeted to chronically ill youth, may become feasible in this context.

Implications for Title V Maternal and Child Health Programs

The Omnibus Budget Reconciliation Act of 1981 fundamentally altered the nature of federally funded maternal and child health programs. The Act established a block grant program to states which incorporated several existing categorical programs, including Crippled Children's Services, which no longer receives a specified federal appropriation. The Act clearly states objectives requiring that states use block grant funds to identify and provide services to handicapped children and youth. As there is no longer a designated state agency under the grant to perform those functions, it is likely that

considerable evolution will occur in the organization of state level services. One option includes the decentralization of public health services to a community level with block grant dollars flowing to local agencies.

This milieu creates a major opportunity for state Title V programs to assess specific needs of children and youth with chronic illness and disability and to respond with innovative programs and services. The Surgeon General's Workshop on Children with Handicaps and Their Families in December 1982 called for several major developments in public health programs (Koop, 1983). The principles elaborated in the recommendations of that workshop have specific implications for serving the needs of youth with chronic conditions.

1. Further definition of the scope of the problem. Despite a broad consensus among knowledgeable program administrators and providers, there is an incomplete data base of the status and outcomes of chronic conditions among youth. Multiple reasons confound the ability to fully define the extent of chronic illness, the primary reason being the lack of any comprehensive morbidity statistics by which these conditions can be monitored over time. Since it is unlikely that such a data base will be initiated on the federal level, states are challenged to ascertain the prevalence of children and youth with chronic conditions within their own boundaries. The task will not be easy, but it is possible.

The care of certain conditions is concentrated among a relatively few tertiary centers in most states, enabling the compilation of demographics on the population served. Third party reimbursement programs, including Medical Assistance (Title XIX) and private health insurers, also have a quantified experience in paying the costs of care for these youth. Voluntary organizations frequently maintain mailing lists or limited registries of individuals in their areas. Special education data provide accurate counts of youth with select conditions who receive services under the Education for All Handicapped

Children Act. Other agencies, including Supplemental Security Income (SSI), Vocational Rehabilitation, and Developmental Disabilities may make further contributions to the data base. A grasp of the scope of the problem in any specific state requires this type of data in order to assemble a needs assessment relating to the complex social, health, educational, and family-related needs of children with chronic conditions.

2. Development of model standards for care. Only selected chronic conditions in children, represented by congenital heart disease, have received careful scrutiny in terms of requirements for care. The identification, evaluation, and provision of coordinated services at all levels of medical care must be assured. Title V agencies, especially Crippled Children's Services programs, can provide a major stimulus for this work, especially if they seek the cooperation of concerned professionals and organizations. Without directly attempting to regulate selected services as a means of compelling desired care models, these agencies can create an expectation for services which quickly become the standard in the community. Several examples exist of the success of this strategy in the past, including the organization of multi-disciplinary care for children and youth with cleft lip and palate, myelodysplasia, and hemophilia. The increasing effort of the marketplace and determination of the scope of care can be positively influenced if public and health care professionals share some major expectations regarding these youth.

One area of inevitable, and necessary, collaboration will be the joint work of health care providers, educators, and those agency professionals who have the responsibility for vocational preparation and placement. The lack of a continuum from home to school and into the work place is a serious impediment for many youth with chronic conditions. The responsibility "boundaries" of agencies currently create disruption and fragmentation of opportunity. This situation seriously compromises a standard of service that is necessary

to assure maximal adjustment of the youth with chronic illness or disability. We have made little progress if our only accomplishment is the control of health symptoms when the individual with the chronic condition does not have the skills or chance to obtain gainful employment and economic independence. Title V agencies can ill-afford their current emphasis on the needs of young children if those efforts dissipate during the adolescent years.

3. Encouragement of systems of regionalized care. While no simple regionalization of services is either politically or physically possible at the present time, considerable duplication and fragmentation of medical care can be avoided if centers and providers offering exemplary service assume a major role in the health care of youth with chronic conditions. Title V programs now have the opportunity to stimulate and encourage exemplary services through grants and contracts that nurture the comprehensive, interdisciplinary approach to care. Rather than diluting a relatively small resource by providing fee-for-service reimbursement for a small number of youth, these funds might now be more appropriately expended to support a system of services not readily reimbursed by third parties on an individual patient basis. The commitment to interdisciplinary care implies that providers, clinics and hospitals will mobilize necessary professional staff to meet complex needs of families. At the present time, many hospital-based programs are finding it increasingly difficult to maintain such staff with patient revenues. By recognizing excellence and competence, Title V agencies can foster the maintenance of these programs through grants and contracts, assuring accountability for the use of funds. In this context it is possible to fund certain services on a regional basis without the onerous regulatory function of denying other centers the "right" to provide the service. Such regionalization does not exclude or diminish the primary health provider or secondary center from full participation in the care of youth with chronic

conditions. Most youth with low-morbidity problems will in fact continue to receive the vast majority of their services from these resources. Regionalization will only occur as a logical means for provision of highly technical or complex services.

4. Creation of alternative financing mechanisms. Despite many extensions and alterations of basic health care financing, comprehensive reimbursement for services remains centered on procedure-oriented medical care and those services which are provided during an acute hospitalization. Crippled Children's Services programs that provide reimbursement for such care have, for many years, established limitations on the use of their funds due to the small Title V appropriations and burgeoning costs of such services. The inevitable result of this policy has been that Title V resources are used for relatively fewer and fewer youth in restricted settings. Even more ominously, administrative decisions have sometimes been based on medical diagnoses, generally excluding children with diagnoses not considered as severe or potentially disabling as other conditions. Therefore, from a national perspective, there is no baseline set of services available to all youth with chronic conditions assured by Title V. Some states continue a constant struggle to meaningfully finance health care for a segment of the target population, but the unmistakable trend is to more restricted coverage.

Other approaches to financing comprehensive services must be sought, especially for youth in families with modest incomes. Whenever possible, resources should be shifted from the highest cost setting, generally an acute care hospital, to a lower cost community service when medically prudent for the chronically ill youth. Such situations, however, represent only a small fraction of the youth with chronic conditions. It would appear more logical that Title V agencies expend considerable professional effort in the immediate future to assure that major reimbursement resources, especially Medical Assis-

tance, health maintenance organizations, and commercial health insurers, meet the comprehensive needs of chronically ill and disabled youth. Frequently, administrative decisions are made by these payers on short-term cost containment. They must be convinced not only that the outcome for their clients will improve, but also that their own agencies, organizations, or companies will benefit in the short-term by a broad-based reimbursement policy. Many Title V agencies have the experience in provision of services over many years to selected populations of children and youth that could illustrate this very issue. There is, perhaps, no more critical area demanding leadership from Crippled Children's Services programs. Should our health care system evolve so that it is generally impossible to fund comprehensive service because reimbursement is based upon acute care requirements, chronically ill youth will suffer a severe setback in the evolution of their care.

The primary expectation of Title V agencies should be that their leaders view the present time as an era of flexibility and opportunity rather than one of retrenchment and defensiveness. Since its inception, Title V programs have made significant contributions to the prevention and management of chronic conditions in children and youth. Models of care that may have been relevant during their initiation must be reassessed. Priorities that appeared logical in another time must be reassessed. The agency role in a spectrum of complex public, voluntary, and private services must be reassessed. What emerges from such a process can be a vital, contributing public health entity that will positively alter the integration of chronically ill youth into society.

Implications for University Affiliated Facilities (UAFs)

The requirement for innovation presents a particular opportunity for University Affiliated Facilities that have experience in providing exemplary interdisciplinary services and training. Team models of professional inter-

action have been most highly refined in settings serving individuals with developmental disabilities. In recent years similar models have been implemented in other chronic disease care settings. Nevertheless much chronic care remains fragmented and incomplete.

Recognizing the behavioral and developmental sequelae of most chronic diseases, it appears appropriate that University Affiliated Programs expand their activities in these areas. Involvement can include direct service as well as training of professionals and clinical staff serving these youth in other settings.

The developmental perspective of adolescence must be incorporated into programs and services working with youth with chronic conditions. Primary UAF training and continuing educational activities can emphasize this perspective and help providers to bridge the transition from childhood to adult life. Aspects of that transition, including transfer into an adult health care service system, are traumatic and sometime counterproductive unless the youth, family, and providers are well prepared.

The growing interest in the adult service system among UAF staff enables these programs to facilitate many of these changes. UAFs possess valuable experience and information which can be used in model development. Whether from the perspective of a specific professional discipline, or from an interdisciplinary team, UAFs must become involved in planning for services to youth with chronic illness or disability.

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METHODOLOGY FOR FORECASTING SERVICE NEEDS FOR
CHRONICALLY ILL AND DEVELOPMENTALLY HANDICAPPED PERSONS

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Design and maintenance of human service delivery systems requires a mapping of the service needs of clients to availability of those services and a periodic assessment of gaps that exist between the two. The principles that govern this process within health care planning domains have been well articulated and accepted worldwide through the collective efforts of the World Health Organization (WHO) over the past 30 years. Many countries have adopted WHO policies with regard to regionalization of health care service and manpower planning in order to improve the quality, distribution and utilization of services, and the implementation of programs in prevention and disease control not otherwise possible. The United States, too, has slowly moved in that direction and instituted a nationwide network of Health System Agencies (HSAs) in the second half of the 1970s with the legislative mandate to monitor availability and accessibility of services. Substantial portions of that system, however, have by now been dismantled.

In contrast to this comprehensive approach to health care planning, the field of service provision to chronically ill, physically handicapped and developmentally disabled persons evinces a much less organized effort toward monitoring service needs and how they are being met. The recent health policy studies by Hobbs, Perrin, Ireys, Moynihan and Shayne (1983) have again documented that there remain major gaps in services to children who suffer from the eleven most common chronic illnesses of childhood and that a systems

approach is indicated to ameliorate the deficiencies. Similarly, the partnership between the private voluntary sector and the Division of Maternal and Child Health, U.S. Department of Health and Human Services, toward regionalization of neonatal care and genetic services during the past decade has created enormous opportunities for habilitative referrals for newborns at risk for developmental disorders. However, there has been a lag in planning for service needs of these newborns by the educational-habilitative network (Davidson, Reif, Shapiro, Griffith, Shapiro & Crocker, 1984).

We are not without models for planning in the field of chronic illness and disability (Cohen & Kligler, 1980); however, few detailed outlines of methods for forecasting service needs are available. Hence, efforts toward this essential component of the planning process tend to be idiosyncratic. For example, a few states have instituted comprehensive data collection procedures on developmentally disabled clients who receive state services. The Child Find program of P.L. 94-142 was intended to create a database documentation of the number of children who would require special educational services, an effort that was both age- and time-limited. Most states maintain a county-based reporting system for providers of generic developmental disabilities services, genetic services, and services to physically handicapped children, principally used to develop mandated statewide plans for service. In addition, there are a small number of examples of comprehensive regional needs assessments as preliminary steps in the implementation of new services and programs for persons with developmental disabilities (Davidson & Crocker, 1977).

The purpose of this paper is to review problems in forecasting needs for health and allied health care services to children and adults with chronic physical and/or intellectual handicaps. The intent is to present a conceptual framework for such needs assessments and to provide potential users with

examples of methodologies to assist them in implementing assessment at the local or regional level.

Conceptual Issues Relating to Forecasting Service Needs

Static versus Dynamic Need

No human need, when measured across a population or subgroup of population, can be said to be static with regard to health and human services. The very nature of treatment and prevention services is to alter the need for those services. Even without designed intervention, human needs vary with a large number of factors, both biologically and environmentally determined. Hence, service needs must be viewed as dynamic. This proposition directly defines the requirement for systematic and periodic monitoring of needs. For example, most federally mandated health and human service plans at the state and local level are redeveloped every three years. The ideal timetable for periodic needs assessments, therefore, would coincide with these existing requirements. Additionally, goals for service provision indicated by needs assessments should be operationally stated and monitored at least annually at the local level.

Relation of Incidence and Prevalence to Need Determination

Among the most widely used indicators of need for health care services are estimates of incidence (the number of new cases of a disease per 1000 population per unit of time) and prevalence (the number of cases per 1000 population at a given point in time). Estimating the incidence and prevalence of definable clinical entities, as for many chronic illnesses, can be accomplished with considerable precision. Gortmaker's (in press) efforts that present incidence and prevalence trends in eleven chronic illnesses stands as an excellent example. However, not all developmentally disabling conditions share such precise definitions. For example, there is considerable varia-

bility in population estimates of mental retardation, the most common of the developmental disabilities, stemming in part from lack of common definitions and less than adequate prevalence estimating methodologies. Despite the variation, prevalence data can and should be applied for planning within states where common definitions are usually applied for statewide reporting.

Level and Scope of Need Determination

Monitoring and planning human services should occur at three levels: policy, strategic, and operational. No such hierarchical planning system is in place at the federal level in the United States, due in large degree to a bifurcation of responsibility for services and manpower development among numerous federal agencies. A similar situation exists at the state level. As a result, planning systems have evolved in idiosyncratic fashion, with varying scope and at varying levels of government. Moreover, most planning is conducted by agencies which also are responsible for provision of direct service. The interests of the agency may not be identical to the interests of the community served.

In addition, manpower planning, an essential ingredient in overall health care planning, has not been comprehensively addressed at either the state or the federal level in the area of chronic handicaps. While the Division of Maternal and Child Health, the Administration on Developmental Disabilities, the Office of Special Education Programs, and the Office of Vocational Rehabilitation have all addressed issues of training of professionals to enter service careers in behalf of handicapped persons, none of these efforts has been consistently based on empirical indices of need.

Ideally, the forecasting of needs for services must employ methodologies that tap both service and manpower needs simultaneously (Mejia & Fülöp, 1978), sampling such data as perceived needs for services by the consumers, services utilization trends, manpower supply and training needs, and demographic and

economic information. The unmet need will be defined by the difference between the actual need and the utilization of available services. Moreover, the planning context, in an ideal case, should occur out of a systems model, where integration and services context can be responsive to the data gathered (Hyman, 1976). The only mechanism in place in the United States that comes close to this level and scope is the individual states' Developmental Disabilities Planning Councils. These interagency planning bodies have the representation and mandate within most states to function at near the ideal level to implement systematic planning, but do so only in very few cases.

Planning and Public Policy

So long as the financing of human services to persons with chronic handicaps resides primarily within the federal and state governments, the planning of services to meet needs will be tempered by what is politically or financially possible (Hobbs et al. 1983). Numerous examples of obstacles to service planning and implementation that stem from this issue can be cited. The financing of services to persons in residential facilities has derived largely from Title XIX, forcing most states to develop community based alternatives to institutional services through other means. Recent amendments to the Title XIX regulations permitting the development of Intermediate Care Facilities for the Mentally Retarded (ICF-MR) have accelerated the pace of community residence development but have not kept up with the need. In this instance, the planners in many states knew the need, but were unable to implement change in services because financing was not available.

A second example is the determination of service catchment areas by means of geopolitical boundaries and not service needs. While techniques exist for empirical determination of catchment areas and have been widely used to determine health care regions, the determination of eligibility for most human service systems for handicapped persons has been governed by such factors as

county of residence or income level. One geographic location may provide comprehensive care and another, insufficient services, to two individuals with exactly the same disabling condition. Such situations prompted the conclusion by the Rand Corporation (Kakalick, Brewer, Dougharty, Fleischauer, Genensky & Walker, 1974) that some mechanism should be added to human service delivery systems that would direct clients and their families through the complexities of locating appropriate services--a seeming paradox of sorts.

Recognizing that some of the issues we have raised so far may not be circumventable for planners of human services, we implore the reader to at least adopt Wolfensberger's (1972) charge to identify wretched compromises. With that caveat, we present some models for forecasting services that have worked within other contexts.

Traditional Approaches to Need Determination

The many approaches to need determination reflect the many different health policy approaches. Agreement exists only on the abstract level of general goals; rarely do we find agreement on how to reach those goals. We have to project needs into the future and plan ahead to meet these needs, but who should do the planning? How should the services be organized? Who should fund them? Health and habilitation are difficult to assess and monitor, existing data systems are full of measurement problems, and crude criterion indicators like infant mortality and developmental outcome are usually not sensitive enough to assess changing needs or to plan services. For example, how does one interpret infant mortality? Should one celebrate the all-time low of our current infant mortality as evidence for the effectiveness of our health care system or deplore its absolute level as shockingly high in international comparison? Similarly, how does one evaluate the differential effectiveness of alternative approaches to habilitative interventions? Never-

theless, some basic planning techniques and data systems are available to aid rational decision-making. Most models have been developed around monitoring of child health systems, however, these models can be adopted for use with other populations.

Demographic Models for Need Projections

Life table analysis in combination with vital statistics recording and a regular census of the population provides the most powerful methodology to project population trends for decades ahead. For example, the American population is currently undergoing rapid changes with crucial implications for pediatrics. The proportion of children in the total population has decreased from 35.7% in 1960 to 27.6% in 1981. Although the total population increased during that 20 year period by nearly 50 million, the number of children (under 18 years) decreased from 64.5 million to 63.2 million and will continue to decrease in the future. Census Bureau data show that our total fertility rate has fluctuated around 1.8 children per woman since 1972, well under the "replacement level" of 2.1, and is unlikely to rise again in the near future.

The number of chronically ill children will decline accordingly. There has been no evidence for any changes in incidence of the major chronic illnesses, although the promise of genetic screening and early prenatal diagnosis will probably lead to a significant reduction for some specific diseases in the future. Table 1 gives incidence rates for ten common conditions that can lead to chronic illness.

Table 1

Incidence of Ten Chronic Illness Conditions

<u>Disease</u>	<u>Cases per 1,000 Live Births</u>
Chronic Kidney Disease	2.0
Trisomy 21	1.5
Spina Bifida	1.0
Cystic Fibrosis	.5
Sickle Cell	.4

Juvenile Diabetes	.4
Hemophilia	.13
Muscular Dystrophy	.14
Leukemia	.03
Phenylketonuria (PKU)	.04

Using population projections for given service areas and these incidence rates, one can make fairly reliable projections of service needs.

The greatest advances in biotechnology occur now in the field of genetics and perinatal care; the psychosocial treatment approach currently dominates the later years, especially adolescents' health care. Indeed, children were never as healthy as they are now. At the same time, we probably never had such uneven distribution of health care needs as now: the great majority of children are in excellent health, but a minority of children with special needs has emerged to claim an ever increasing share of all child health expenses. During the first year of life, some 4% of infants needing intensive care account for 90% of all inpatient care. A similar, but not quite so dramatic, clustering of health care needs continues in the later years of life. Ironically, much of the new need is due to advances in medicine. Children with minimal survival chances in the past now live into young adulthood. Advances in neonatology improved the newborn care for all, but the effect is most pronounced for those born prematurely and at highest risk for developmental delays (Budetti, Barrand, McManus & Heinen, 1981). Despite dramatic declines in mortality in this population over the past 20 years, however, there has been a slight increase in morbidity associated with perinatal illness. This trend bears direct monitoring in the future since it is likely to continue and to vary with better health care and better early rehabilitative interventions.

Data Collection to Monitor Child Health

Most European countries have a secretariate or ministry for family and youth which monitors maternal and child health and administers programs of

health and social services for mothers and children. No equivalent institution was ever developed in this country. Data on children are gathered as part of the regular activities of the Bureau of the Census or of the National Center for Health Statistics (NCHS) at the Department of Health and Human Services. The National Health Interview Survey, the Health Examination Survey, the National Nutrition Survey, and the Hospital Discharge and National Ambulatory Medical Care Survey all include children, but reports focusing specifically on the health of children are only infrequently issued. The largest recent compilation of data on child health was prepared for the Select Panel for the Promotion of Child Health (DHHS, 1981). A number of recent conferences have been held to address issues of monitoring child health to provide data for public policy evaluation (Klerman, 1982). Data systems have been established by children's advocacy groups, by representatives of state agencies and by professional organizations. None of these appear as yet completely satisfactory; however, some may be useful to states in developing monitoring systems.

Monitoring the Health of Children with Special Needs

Given the deficient data base for assessing and monitoring child health in general, it should be no surprise that population data on chronic illness and developmental handicaps are even more deficient. Even the National Health Interview Survey (HIS) with data on some 45,000 families nationwide does not cover enough children to monitor the rare diseases that have a prevalence of less than 1 per 1000. The HIS has, however, one general question concerning limitation of activity due to "chronic conditions." In 1980, some 3.8% of children age 0-16 had some activity limitation due to chronic conditions reported, including 2.0% with a limitation in major activity. This is consistent with other estimates of chronic physical conditions of 1-2% (Hobbs et al. 1983). Projections such as this, combined with the incidence rates given

previously for specific chronic illnesses, can be employed to project needs for chronically handicapped children.

Needs assessment for rare chronic physical conditions and major developmental delays necessitates modification of methodology used in general health monitoring. The needs assessment and the service components cannot easily be separated. Because of the high direct and indirect costs to the family, some public support for special programs is usually required, creating a multitude of organizational problems for coordinating private and public services. The politics of public funding for professional services rarely allow optimal solutions. Methodologically the implications are that original data collection using random sampling approaches in the population are inefficient; planners should employ "snowball sampling" and use data from existing files, such as charts in private practice, follow-up programs for high risk infants, and school readiness tests. Surveys of identified cases in specialty clinics at medical centers, referral and directional services centers, and birth defects and chronic or genetic disease registers can provide surveillance and allow optimal service planning.

Current Planning Efforts

Political and historical factors have led to a pluralistic health care system under strong provider influence (Starr, 1982) with only two major planning directions. The first one deals with the political regulation of health care establishments. The second direction concerns the internal planning of voluntary services and the administration of public monies allocated to services for the poor or for categorical programs.

An example of the first kind of planning efforts has occurred in the Comprehensive Health Planning (CHP) agencies as called for in P.L. 89-749, funded by the federal government through grants to states since 1967 and later replaced by the National Health Planning and Resource Development Act

(P.L. 93-641) of 1974. This act created approximately 200 Health Systems Agencies (HSAs) with a mandate to develop regionalized health plans, to review proposals for new services and to prepare recommendations to their state certificate-of-need (CON) agencies and State Health Planning and Development Agencies. HSA staff prepared need assessments, obtained community input, and made recommendations to the HSA council.

Extensive planning methodology was developed in the second half of the 1970s to provide population and resource data for detailed geographic areas. Typical for this methodology are the Area Resource Files (ARFs) which contain merged census and service files to provide need and service profiles for all HSAs and counties in the country. Similar files were prepared within HSAs to describe census tracts and neighborhoods. Need documentation was required to obtain federal funds for new health centers or new service programs. With the institution of cost containment efforts in 1980, most of the regulatory mechanisms to back up such health planning activities were dismantled and replaced by competitive mechanisms and reimbursement limits, although many states and local communities still employ the methodology.

Even with the diminished regulatory mechanisms, there remained the second planning effort. Public programs for the handicapped (Title V) and for the poor (Title XIX) had to be administered. Local health departments and social service departments have to coordinate their services with state departments to administer the Maternal and Child Health block grants. State Developmental Disabilities Planning Councils publish plans that integrate numerous state agency plans for resource allocation. School districts must comply with the legislation on Education of the Handicapped (P.L. 94-142) and plan accordingly. Community agencies funded through the Community Chest and other voluntary efforts need to state objectives and to plan and coordinate their services.

Though local power processes and political bargaining are finally more influential in resource allocation than documented needs assessments, goals have to be clearly stated and needs have to be specified as input to the political debate. Whenever the free market regulation of services through demand and supply is not applicable, planning mechanisms have to take its place. Honest persons may disagree on how best to organize human services, but they do agree that some organization is needed to provide services.

Some Examples of Forecasting Needs

Specific publications dealing with service needs are rare. There are some discussions of national plans like that of Talbott (1981) and reports of a number of national level commissions and select panels. Manpower projections based on national need requirements have been provided for various disciplines as in Roddy's (1981) report relative to primary care physicians. Such national manpower projections use age-specific incidence and prevalence rates for acute and chronic disease, population projections and recommended visit schedules based on official standards for good medical care.

State and local need and service projections exist widely but rarely find their way into the scientific literature. The interested audiences are too small and localized and can easily be reached through special mailing lists. The authors are demographers, health planners, or survey researchers working in non-academic settings where publications may be neither encouraged nor rewarded. Local and state planning offices and research agencies and private consulting firms prepare such reports on request or under contract for specific purposes. The cost of preparation and printing, combined with small interested audiences, discourages their wide availability.

Nevertheless, some studies of localities or specific facilities reach the scientific literature to draw our attention to current deficiencies. One

example is the by now well-documented gap in primary care services for chronically sick children. Stein, Jessop and Riessman (1983) studied a sample of 209 children with chronic childhood illness seen at a municipal hospital in the Bronx, using bilingual home interviews. Carroll et al. (1983) surveyed 61 adolescents visiting six specialty clinics at a large children's hospital in Montreal, using a self-administered questionnaire to be completed by the patient at a scheduled visit. Palfrey, Levy and Gilbert (1980) interviewed 158 parents bringing their children to five specialty clinics at Children's Hospital in Boston. All of these researchers identified serious gaps in the provision of primary care to chronically sick children and, often, the complete absence of sources of care other than the specialty clinics. The methodology usually involved small samples and personal interviews or self-administered questionnaires completed at the place of care during a relatively short period, e.g., one month. The analysis required only simple frequency distributions.

Another easily documented need is for support networks among parents with mentally retarded children. Eheart and Ciccone (1982) studied the needs of 36 low-income mothers in the first year after learning that they had a retarded child. Directors of child care centers identified subjects for the investigators and allowed a 45-minute interview at their center. There was an overwhelming need to learn coping skills, suggesting the establishment of support groups together with early intervention treatments for the children. Another study (Suelzle and Keenan, 1981) of support networks over the life cycle of retarded persons used a mail questionnaire to obtain data on 330 parents with children ranging in age from preschool to young adult status. Younger parents made more use of available services and were more supportive of mainstreaming. Parents with older children tended to be isolated and in greatest need of expanded services.

The provision of services in sparsely populated rural areas is always problematic, but poses especially difficult problems for the rehabilitation of chronically sick people. Omohundro, Schneider, Marr and Grannemann (1983) studied four rural counties in Texas, Oklahoma and Arkansas, using local interviewers to identify and visit 456 respondents, identified from county lists of people with known handicaps. Identified needs were limited mobility, limited employment opportunities, difficulties with house and yard work and multiple physical and emotional health problems. Few of the needs were addressed in any one of the counties. Sample sizes of just over 100 per county were sufficient to provide a reasonably clear picture.

Finally, lack of adequate educational services for handicapped preschoolers was studied by O'Connell (1983). She used a small Faculty Contingency Fund of her university to mail to all directors of teacher certification in the 50 states and the District of Columbia a one-page questionnaire to ascertain which states have mandated services for handicapped preschoolers and what teacher certification is required for these services. After a second mailing and telephone follow-up, information from all states and the District of Columbia was obtained. Only 16% of the states reported that services are mandated to all handicapped preschoolers. Several strategies for change are advocated: Development of active parent organizations, education of the general public, linkage between educational personnel and child-care professionals, and the formation of legislative task forces to contact legislators prior to any relevant state legislation. A very moderate research investment had provided a wealth of important information.

In 1977, the Developmental Disabilities Office (now the Administration on Developmental Disabilities) authorized an expansion of the UAF network through the creation of Satellite Centers. Five such centers were developed as extensions of the existing UAFs based on the results of feasibility studies. The

intent of each feasibility study was to establish the need for new services, manpower development, technical assistance, and dissemination activities related to developmental disabilities in areas of the U. S. where such services had not been available. Studies took place in Hawaii, Montana, Vermont, Western New York, Western North Carolina, Minnesota, and the Navajo reservation centered in Tsaile, Arizona, each funded with about \$25,000, and completed in about six months.

Each study required the collection of empirical data specific to the identified catchment area describing (a) the prevalence of developmental disabilities and characteristics of the known developmentally disabled population (obtained from existing data in state plans for services), (b) the numbers of developmentally disabled persons receiving direct and support services (obtained by agency and direct client surveys), and (c) the characteristics of the developmental disabilities service delivery system and its current capacity to provide direct and support services (obtained by agency survey). These data allowed study teams to specify the unmet needs of the developmentally disabled population and the barriers to meeting those needs from the viewpoint of both consumers and service providers.

All studies assessed needs for services and manpower development simultaneously. Methods of data collection varied somewhat from study to study, however, all study teams employed direct survey techniques involving both mail questionnaires and face-to-face interviews of consumers and agency staff. Also, all teams employed variants of a delphi technique to achieve consensus among service providers, consumers, representatives from higher education and government officials relative to unmet needs.

Additional satellite studies were funded in 1984, for which a new format for conducting studies has been written (Davidson & Fifield, 1984). The technology demonstrated in 1977, and continued for 1984, is geographically

nonspecific, comprehensive, and generalizable to any disability, service delivery system, or geopolitical entity. It is a relatively inexpensive process that is not particularly manpower-demanding, as much of the effort must be placed in gathering existing data. It does require a minimum of six months to adequately complete direct surveys which, in our experience, are of inestimable value in validating conclusions.

The few studies cited give a flavor for the ongoing research. Small samples, carefully selected from lists of clients, providers, or government officials, are approached with questionnaires or interview schedules, to be analyzed with simple descriptive statistics. No fancy designs are needed; no sophisticated measurements are involved. Service gaps are easily documented, but solutions are difficult to find. They often require new resource allocations that can only be achieved through the political process. Lists of local providers, such as primary care physicians, educators, school nurses, social workers and voluntary agency staff are needed for sampling and frequent polling. Constant contact and coordination, through mailings or committee work, with the provider community is required to continually reassess needs and services.

Conclusions and Implications for University Affiliated

Facilities and Title V Programs

The current pressure on governments and service providers alike to contain costs coupled with recent budgetary constraints in human services must force us to carefully examine alternative strategies to meet the needs of our clients. While there have been ample reasons for the field of developmental handicaps to implement and maintain an enduring means for assessing service and manpower needs, the current pressures make such monitoring essential. Only by carefully monitoring needs can orderly change in service delivery be

justified and resources be efficiently applied to meet the changing need.

The review we have just undertaken leads to several conclusions. First, forecasting the needs must become a regular part of human service systems. Second, the changing needs of developmentally handicapped people must be monitored across all systems charged with provision of services to them. It is thus necessary to explore strategies that will effect cross-system integration. Third, needs for services and needs for manpower must be monitored simultaneously, employing operational definitions for each that are understood and commonly accepted by both service providers and manpower developers alike. For instance, it is ineffective for a human service agency to specify a need for a care provider with a specific level of training when there is no realistic chance of finding or funding professionals with those credentials. A better strategy would be to use manpower needs assessments to influence the distribution of discretionary funding toward preservice education. Finally, needs forecasting must influence operational, strategic and policy planning levels simultaneously. Neglect of this principle could result in either policy change with no implementation capability (as has happened repeatedly in health and education systems serving handicapped persons) or attempts to implement services with no chance of permanent funding or regulatory maintenance.

It is acknowledged that the introduction of needs forecasting to human services that each year overspend their already condensed budgets is a difficult task indeed. As resources with government agencies shrink, planning and monitoring functions often give way to direct service provision. This script has been played many times over within state Title V programs during the past few years. It may be necessary for states to turn to other resources for assistance in these functions. Many UAFs have the expertise to design monitoring systems; some have already performed such functions within their

states. The UAFs also are often the only agency within their states with such expertise and with linkages that cut across the health, human services and educational systems, and manpower development programs.

The UAFs as a group rarely, if ever, undertake forecasting of market needs for personnel, the very mission that those agencies are funded to accomplish. In an era of fiscal constraint, it behooves training agencies to understand the needs of the market and prepare professionals to match those market needs. Performance of training in the absence of such data represents a substantial risk for the training agency and to the service community and consumer as well.

Thus, there are good reasons for both Title V agencies and UAFs to share resources to develop and implement forecasting systems within their states. These systems may initially be costly but will in the long run yield better matches between need, suitable services, and available manpower, and will therefore prove to be cost effective. The use of existing resources should also enhance the cost efficiency of the effort.

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REGIONALIZATION OF HEALTH SERVICE SYSTEMS

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The concept of "regionalization," as applied to systems for provision of health care services, refers to the process of active planning for the coordination of activities and resources across a geographic area. As Ginzberg (1977) states in his definitive volume on this subject, regionalization describes programs "that alter the functions of--and relationships among--health providers within an area in order to achieve better access to health care, a higher quality of service, lower costs, greater equity, and more responsiveness to consumer needs and desires." He further comments that "regionalization implies a form of resource allocation or service delivery rooted in geography."

The breadth of geography or population involved in the planning is not critical for this discussion--it could be within a city, state, region, or even a country. The key matter is that consideration is being proactively given to the needs of a larger cluster of potential consumers to achieve improved effectiveness.

Being involved in a regionalization system can imply for the provider some diminution of personal freedom, but the alliances formed and the pooling of resources carry a compensatory excitement. When the mission is better defined, the strengths of many centers brought together, and a diverse population aggregated, there can be expected an ultimate enhancement of the options for both those imparting service and those receiving it. One can anticipate that for the region under discussion there will be:

- a. broader access to advanced knowledge and technology

- b. more people served, and more energy expended in recruitment for use of services
- c. achievement of a critical mass of clients in the area of low-incidence problems so that situations are better understood
- d. avoidance of duplication of, or competition for, incompletely utilized services
- e. funds available which will allow personnel for coordination, data collection, epidemiologic study, research and training
- f. achievement of credibility for negotiation of new payment plans, enforcement of quality control, procurement of grants, and political action.

In a program of regionalization, there is obviously a requirement to have a headquarters wherein policy determinations are secured and certain key central functions accomplished. Hence the nomenclature of "centralization" can be applied to some aspects of the regionalization phenomenon. It can be said that a region is served from the administration of a central base, with outreach. The paradigm is that of diffuse/organized vs. diffuse/unorganized services. There is obviously a strong obligation to ensure participatory governance which will avoid stultifying control; such challenge is paramount to the lasting success of a regional program.

One model of centralization which has served as a prototype for the advantage from pooling is that of biochemical analyses of specimens from newborn infants regarding the possible presence of phenylketonuria or congenital hypothyroidism. In these low-incidence situations (1:12,000 and 1:4500 births, respectively), laboratories handling a small volume of samples are not able to provide detailed technical attention to the analytic vagaries or the programmatic support needed to offer interpretive assistance and follow-through. Virtually all instances of "missed" diagnoses have resulted

from utilization of collection systems and laboratories with less than full-time investment in these demanding studies. As a result of the practical (and legal) pressures to involve the efficiencies of centralized laboratories, the random use of small private labs for phenylalanine or T₄ analysis has virtually disappeared. In the vast majority of circumstances large state laboratories are employed. In four areas of the U.S. specific regional programs have been formed: states of the Northwest, the Rocky Mountains, the Mid-Atlantic area, and New England. In the latter region (based at the State Laboratory Institute in Massachusetts), there is a pooling of specimens from 4 states (MA, NH, ME, RI) for PKU detection and 5 states (MA, NH, ME, RI, and CT) for hypothyroidism screening (Levy, 1984).

To exemplify the various designs and goals of regionalized programs, four representative illustrations will be given:

- I. Genetic services in the state of Ohio -- an instance of a state systematizing its considerable assets in genetics
- II. Massachusetts Maternity and Newborn Regionalization Project -- creative planning for improved care of newborn infants
- III. New England Regional Infant Cardiac Program--a confederation of professional colleagues to achieve better management of a compelling though low-incidence clinical problem
- IV. New England Regional Genetics Group -- an alliance of service providers, public health planners, and consumers.

I. REGIONALIZATION OF GENETIC SERVICES IN OHIO

Based on Eaton, Peppe, Quilty, Peterson, and Ackerman (1982) and Eaton, Ackerman, Linyear, and Sfakianaki (1978)

A regionalized system of health care delivery for genetic services in Ohio was formulated and implemented in 1976. The purpose of the statewide university-based network was initially to prevent mental retardation and other

developmental disabilities, with the long term hoped-for benefit being prevention of other genetic disorders having serious consequences. The Ohio Department of Health assumed the lead agency role and administers the project. The program involves six genetic centers located within hospitals and medical schools (Cleveland, Columbus, Cincinnati, Toledo, Akron, and Dayton), each serving a potential population of 2 million persons. In addition, three satellite clinics are located in more remote geographic areas and served by regional center staffs.

The principal objectives of the Genetic State Plan are (1) identifying certain selected genetic and/or metabolic disorders in infants through screening programs, (2) identifying mothers and families at high risk for genetic disorders, (3) providing public and professional information and education, and (4) developing a centralized information system. The genetic centers provide a comprehensive range of clinical and laboratory services, professional and public education, and are also involved in research. At the state level, screening for newborn metabolic and endocrine diseases is conducted through a centralized state health department laboratory. An advisory body with professional and consumer representation has been established. Coordination of the program is accomplished through regular meetings of all center directors under the sponsorship of the Ohio Department of Health.

Funding for this service network is derived from a variety of sources, including:

- a. federal appropriations through Title V, via the Maternal and Child Health Block Grant
- b. state funds for the genetic services network appropriated by the Ohio Legislature as a line item
- c. medical schools and hospitals providing staff and facilities for the centers

- d. private foundations, including the Cleveland Foundation which in the past assisted the Cleveland Regional Genetics Center
- e. contributions from voluntary organizations, most notably the March of Dimes Birth Defects Foundation
- f. the Ohio Developmental Disabilities Planning Council, which contributed funds for the pilot Hypothyroid Screening Program for Newborns and funded a public awareness campaign in genetics, and
- g. the Ohio Department of Mental Retardation and Developmental Disabilities which supported a cytogenetics laboratory within one state institution for mentally retarded persons.

Support for this program grew from \$150,000 in 1976 to \$1.3 million in 1982. There has been a major shift from federal dollars to state funds. The statewide Genetics Network is now established on a permanent basis in Ohio law. Client payments are 59% from private insurance, Medicaid 10%, and self-pay 10%. The total caseload served from 1979 to 1981 was 29,465 persons.

A strong emphasis on public and professional education is evidenced by the significant number of individuals reached in both categories (in excess of 33,000 contacts for professional education and over 20,000 consumers). Examples of the former include medical school modules, nursing school lectures, continuing education and newsletters, and of the latter, Junior/ Senior high school lectures, a film library, health fairs, and mass media.

In summary, the Ohio Department of Health has recognized the importance of genetics to the health professions and as a beneficial preventive public health program by implementing a coordinated delivery system statewide. This unique cooperative venture could well serve as a national model.

II. REGIONALIZATION OF MATERNITY AND NEWBORN CARE IN MASSACHUSETTS

Based on the Final Report of the Department of Public Health, Commonwealth of Massachusetts (1976), Ryan, Pettigrew, Fogerty and Donahue (1977), and Fogerty (1984).

Regionalization of perinatal care can be traced to the development of units for the care of the premature infant in the United States during the 1930s and 1940s, but in that early period there was little in the way of standards for perinatal practice as it is now known and the impact of these facilities on infant mortality in the U.S. was modest (Butterfield, 1983). The first newborn center was established in New Haven in 1960, and there are now nearly 350 intensive care centers. Each has some components of regional outreach, with varying involvement in transport systems and educational programs. Statewide programs for regionalized perinatal care grew rapidly in the early 1970s, with 28 being operational by 1974 (Berger, Gillings & Siegel, 1976). Many national professional organizations (obstetrics, pediatrics, hospitals) gave earnest effort to the formulation of practice methods and standards for evaluation, culminating in the landmark report in 1976, "Toward Improving the Outcome of Pregnancy" (Committee on Perinatal Health, National Foundation).

Massachusetts has had a long tradition of excellence in maternal and child health services. Early leadership was provided by the Perinatal Welfare Committee of the Massachusetts Medical Society, which conducted a major study of perinatal deaths in 1967-68, and proposed a statement in support of "Centralized Community or Regionalized Perinatal Intensive Care," adopted in 1972. The Massachusetts Department of Public Health designated seven neonatal intensive care units as official transfer nurseries in 1970, and was active in that period in the promulgation of basic standards for maternity and newborn care. As a result of these activities, plus the new presence of "Health Planning Agency" units throughout the state, there was a gradual decline in

the numbers of maternity/newborn services in small hospitals where the quantity of deliveries was so low that provision of exceptional care was not feasible. Hospitals offering maternity services decreased from 120 in 1960, to 65 in 1975. Improved care of infants at risk in this period produced a decline in the Infant Mortality Rate (deaths per 1000 live births) from 19.9 in 1968 to 13.9 in 1974.

In an effort to promote further gains in this system, an extraordinary project was carried out in the state from 1974 to 1976, funded by D.H.E.W. and Blue Cross of Massachusetts, to study the potential for enhanced regionalization. With the Department of Public Health as the lead agency, a working alliance was formed among medical specialty groups, universities, hospital associations, health planning agencies, nursing representatives, individual physicians, individual hospitals, United Way, regional federal agencies, and consumer representatives--the Massachusetts Maternity and Newborn Regionalization Project. Visits were made to all maternity services in the state by a team with special expertise consisting of an obstetrician, pediatrician, anesthesiologist, and perinatal nurse. Utilizing a standardized format, all aspects of local clinical and logistical practices were reviewed (referral rates, protocols for high risk deliveries, trained personnel for sick newborns, etc.), and detailed reports of findings and recommendations were returned to the hospitals.

Many objectives were achieved, particularly in the areas of: (a) establishment of communication between high-risk centers and community units, (b) building relationships among the various high-risk centers, (c) promotion of the importance of consultation and referral systems for high-risk mothers prior to delivery, (d) exchange of information on recent advances in maternal and newborn care, and (e) establishment of a model for peer review and ongoing monitoring of perinatal services on a regional basis. During this process

revised standards and regulations were developed for hospital maternity and newborn services, as well as criteria for the functions of perinatal centers. Uniform terminology and reporting forms were developed. New information was gathered on requirements for travel time to special centers, and the relationship between maternity service size, economics, and utilization. The capacity for future health service planning was much refined.

The years around and during the time of the Regionalization Project were important ones in the overall evolution of modern principles for the care of mothers and babies at risk, so that it is difficult to identify the specific effects of that special effort in Massachusetts. Current reflection suggests that the work expedited progress toward establishment of more appropriate transfer of mothers and infants to centers for expert care.

As a result of the Project's consultations, more mothers in difficulty were moved before or during labor, troubled infants were transferred more promptly, and the perinatal centers began to assume responsibility for the transport process itself. The Infant Mortality Rate in Massachusetts fell to 10.4 in 1980 and 9.6 in 1981. Currently there are 59 maternity centers in the state, and practice standards are well supported. Specialized care is provided at 5 perinatal centers (Bay State in Springfield, Worcester Memorial Hospital, and in Boston at Brigham and Women's Hospital, Boston City Hospital, and St. Margaret's Hospital), with additional newborn intensive care units at the Children's Hospital, Massachusetts General Hospital, and Boston Floating Hospital.

III. THE NEW ENGLAND REGIONAL INFANT CARDIAC PROGRAM

Based on Fyler, et al. (1980) and Fyler (1984).

As the 1960s proceeded, major gains in the fields of pediatric cardiology and cardiac surgery produced a substantial improvement in the capacity for

intervention and cure of infants with congenital heart disease. Many of these developments had taken place at the Children's Hospital in Boston, and it was there that a plan was conceived to assist in the availability of modern care for such infants in the six New England states. Discussions were held with the 11 hospitals in Region I where definitive cardiac treatment occurred and an alliance was conceived to share methods, data, and professional education in behalf of this population of patients. Grant support was obtained from the Maternal and Child Health Service in Rockville in April 1969, with the Children's Hospital in Boston designated as headquarters for the Program. The other units were invited to join on a voluntary basis, and all did (Hartford Hospital and St. Francis Hospital in Hartford, CT; Yale-New Haven Hospital in New Haven, CT; Maine Medical Center in Portland, ME; Boston City Hospital, Massachusetts General Hospital, and the Tufts-New England Medical Center in Boston, MA; the Dartmouth-Hitchcock Medical Center in Hanover, NH; the Rhode Island Hospital in Providence, RI; and the University Hospital of Vermont in Burlington, VT).

The specific goal was to improve infant cardiac care throughout New England; in many regards the federation represented the formalization of pre-existing informal relations. The incentives to join were mainly in the area of being included in a positive collegial sharing in a joint search for improved practices, participation in a Region-wide case-finding program (which would enhance referrals from community hospitals for possible surgery), and eventually to have greater eligibility for various federally sponsored follow-up projects. The original grant (about \$150,000 per year) included support for a full-time cardiologist at the Children's Hospital, part-time coverage at Yale and Tufts, a project nurse and a social worker, secretarial help in all units, and computer assistance. Member hospitals agreed to hospitalize all sick infants and to computerize their experience.

The effects of the Program were many. Basic clinical experiences in child management (cardiologic and surgical) were discussed in an academic atmosphere by physicians at regular meetings. A large amount of new knowledge was aggregated and shared regarding the problems associated with various congenital lesions. Nursing education was vigorously pursued on visits to newborn nurseries throughout the Region, resulting in better understanding and referral of the cardiac infant. The needs of families whose infants were suddenly transferred were considered, with some support monies utilized for boarding of parents near the new hospital (\$5-10,000 per year). Transportation practices were standardized, eventually moving toward hospital-based ambulances, with assistance occasionally provided for transport costs (a total of up to \$20,000 per year). Plans for coverage of hospital care costs were developed locally, and existent systems were encouraged to carry out follow-up studies. There were symposia, conferences, and exhibits on the problems of these infants, in professional meetings.

A total of 2251 infants with heart disease were treated in the first six years, 49% receiving surgical intervention. Of particular significance was the development of a 20% increase in the first years of the discovery rate for such infants, with referral into the treatment system, representing a greater realization of clinical responsibility in hospitals of the Region (now known to be 2.6 per 1000 births). Earlier transfer also resulted (30% within the first 48 hours of life). All of the 11 involved medical centers shared in the enhanced patient flow, and the early concern was assuaged that one or two large centers would dominate the referrals. As time went along the Program became a de facto surveillance system, with physicians monitoring themselves on the outcomes of management. Eventually some centers discontinued their commitment to offering medical and surgical cardiac care for infants; at present 5 retain full services, 3 have partial services, 2 provide only medi-

cal services for infants, and one has discontinued pediatric cardiology entirely.

The MCH grants continued at about the same level from 1969 to 1978, with major reduction thereafter, and cessation 3 years ago. Some assistance from state agencies and the Baruch Fund was obtained for a period. Now the Program has ended, although the effects on practice in the Region are still felt. The most significant final outcome of all of the activities in this dynamic field during those years is a change in the survival rate for infants sick with major cardiac disease. In 1969, 50% died by their first birthday, now only 25% do. There are various factors involved, but it seems probable that a critical one is the promotion for the right to prompt and definitive treatment which was the mission of the N.E. Regional Infant Cardiac Program. It is not possible to obtain comparable statistical data elsewhere in the country on such a regional population. It is quite possible that infants with heart disease in Region I enjoy the best survival rate in the world.

IV. THE NEW ENGLAND REGIONAL GENETICS GROUP (NERGG)

Based on Crocker and Tsairis (1978) and Crocker, Erbe, and Kacoyanis (1978a, 1978b, 1979, 1980, and 1981).

NERGG has existed for six years as a consortium of the genetic service providers, public health planners, consumer groups, and federal agency personnel in Region I. It serves as a forum for the discussion of Region-wide concerns regarding provision of optimal service in the genetic disease field. Its unique format has allowed the broad review of genetic issues, formation of special study groups, the calling of relevant conferences, and the creation of continuing projects which assist in service delivery. Most specifically it has built a sense of trust and mutual commitment among the involved workers in this 6-state region, leading to improved systems for care and prevention.

Special planning activities for genetic services in New England began in

mid-1977 with a project contracted by the Bureau of Community Health Services to the Developmental Evaluation Clinic (UAF) at the Children's Hospital in Boston. This resulted in a conference and in preparation of the "Nonantum Report," a consideration of the federal role in genetic service design as exemplified by Title IV, P.L. 94-278 (January 27, 1978). Following this, a continuing program was initiated under the title "Feasibility Study for Enhanced Regionalization of Genetic Services in New England." A "Preliminary Report" (September 27, 1978) surveyed current genetic service activity in the six states of Region I. It was noted that some programs were already operating with a regional design, e.g., services for patients with hemophilia and with retinitis pigmentosa, and analyses of lysosomal enzymes. A meeting was then called (November 30-December 1, 1978) to bring together representatives from the major centers for service programs, public health agencies, and important consumer groups throughout New England for a discussion of regional concerns in genetic services. This session, the first "Windsor-Charles Conference," was held at the New England Center for Continuing Education in Durham, NH.

The Windsor-Charles I conferees unanimously agreed that there was substantial value to be obtained from a sustained regional alliance for genetic service planning in New England. They made the ultimate "regionalization" gesture of forming the "New England Regional Genetics Group," to carry out their intentions, and established seven operating committees to pursue interim study of the major topical areas: Census, Quality Assurance, Prenatal Diagnosis, Education, Third Party Payment, Screening, and Satellite Genetic Services.

A Steering Committee, consisting of the project leadership, a representative service provider and public health planner from each state, and the chairpersons of the operating committees, was assigned to carry on an active

discussion regarding the project's program. This body meets three times each year, with special communications between meetings as needed.

The focal point of NERGG's activities each year has been the Annual Meeting, held in November or December in Durham, NH. A representative attendance is sought from a flexible list based on key workers in the state health departments, the major service centers, strategic special projects, and consumer groups. Experience has shown that some professionals and association persons can be counted on with good regularity, and they constitute the basic "membership" of NERGG; an almost self-selecting adjustment occurs in this as the years pass. The characteristic Annual Meeting attendance includes about 50 persons: 8-10 from state health departments, about 3-5 consumer group representatives, several Regional federal officials, and the remainder from all of the relevant medical service centers of the six states. The diverse qualities of the conferees assure that representative voices will be heard; from this pool of human resources have emerged the major contributors to the Group's actions. At each Annual Meeting there is the opportunity for individual state health departments to report on their own special conditions and hopes, and to share the planning implications with service providers. For example, in 1982 the Massachusetts "Information System for Environmental Effects on Pregnancy" was described. Key speakers from within and outside the Region have highlighted current issues in the genetic disease field.

During the life of NERGG the Operating Committees have contributed the central action. At the formative 1978 session the first seven Operating Committees were established; in 1979 Consumer Concerns, and in 1983 Legislative Liaison, were added to the list. Chairmanships have been stable (with membership somewhat eclectic). The various committees may have conferences and meetings through the year; they are expected to provide a progress report at each Steering Committee meeting and a summary report at

each Annual Meeting. The major areas of Committee interest have been:

- a. Quality Control - discussion of genetic quality control issues, analysis of ultrasound practices in pregnancy, concern with maternal serum alpha-fetoprotein analysis
- b. Outreach - survey of satellite genetic clinics, evaluation of services, identification of underserved areas
- c. Screening - endorsement of current newborn screening programs, consideration of new techniques, use of premarital informational materials
- d. Education - education functions in state genetic programs, Genetic Education Newsletter, field testing of high school genetics curricula, analysis of physician education in genetics
- e. Census - annual review of amniocentesis utilization, consideration of methods for gathering of data on birth defects
- f. Third Party Payment - survey of reimbursement practices re genetic services, contacts with major insurance carriers
- g. Prenatal Diagnosis - affiliation with the 14 diagnostic centers in Region I, monitoring of laboratory utilization, slide circulation for proficiency review, consideration of values in centralized performance of amniocentesis
- h. Consumer Concerns - focus on satisfaction from genetic counseling, voice for interests of self-help groups, exchange with activity areas of other committees
- i. Legislative Liaison - pooling of experiences in the transfer to state government of support for genetic services, analysis of legislative trends with alert to membership as appropriate.

As professional exchange increased within the framework of NERGG, a number of special projects were conceived which gained from the potential for

Region-wide pooling of population and services. Chief among these have been the N.E. Fragile-X Syndrome Collaborative, the N.E. Regional Maternal PKU Project, the N.E. Regional Maternal Serum AFP Screening Program, and the Region I Thalassemia Program. These began as affiliations in common interest, on a voluntary basis. At this time they have all achieved varying levels of federal or foundation support, some of them from several agencies.

NERGG was a pioneer in the process of forming regional alliances for the coordination of genetic services. In the early years DMCH sponsorship was at a level of about \$10,000 annually, covering the costs for the Annual Meeting, some committee activities, travel, and publications. Last year this was increased, to allow support for several strategic regional conferences on key issues in the genetic field and part-time employment of a genetic associate as Coordinator of NERGG activities. It appears likely that in the future the Group can serve as a base for broader support for particular projects. The funding has usually been attached to the DMCH training grant for the UAF at the Children's Hospital in Boston.

The ultimate potential gains achievable for a regional federation of all parties involved in genetic services for a population are in a state of development. Its scope is more abstract or global than the state program described for Ohio. NERGG has recently prepared advisories for health departments and service professionals in Region I regarding practice issues for maternal serum AFP analysis and for chorionic villus biopsy. Regional conferences are now planned on a greater variety of current topics. The transition from federal to state funding for public programs, as the Genetic Diseases Act decelerates and MCH block grants prevail, has been much considered. Advice is being offered to genetic groups in other regions of the country. A sense of exhilaration exists, derived from the excitement of the joining in a deliberate fashion of the many factions in Region I who have a

common stake for improved genetic services.

Comment

It will be apparent from the description of these four projects that in each one most or all of the hoped-for gains from regional planning (a-f, in the introduction) were achieved. There were larger clusters of consumers involved, specialized personnel were brought to bear on uncommon but clinically demanding situations, there was an improvement in the quality of services or coordination, and consolidation of resources was effected where appropriate. Operation of the "centralized" portions of these programs required federal money at the outset in every instance, but at relatively modest levels, and this was followed by an investment of state and private support. Lasting gains in the health service system were produced.

Implications for Title V Programs and University Affiliated Facilities (UAFs)

Directors of MCH/CC programs in state health departments are already involved in many projects of a regional character. In some regards a health department is one large regional program already, since it carries out planned and coordinated services within a geographic area. It is a goal of this paper, however, to point out that other levels of active planning can exist with additional participants beyond the agency which exceed the intensity of "regular" regional operations, that their support can come from a variety of sources, and their results can be important. Directors may wish to analyze the potential opportunities for systems improvement by state-wide projects for key situations where there is low incidence and/or new technology (as with the Ohio genetics program or the Massachusetts maternity and newborn service study). Further, they may wish to consider linkage with other states in a special laboratory assignment (such as the New England Regional Newborn

Screening Program). Beyond this they can be supporters or involvees in innovative multi-state service or coordination projects where fragmentation existed before, e.g., the New England Regional Infant Cardiac Program or NERGG. Which portions of the maternal and child health systems in the states could most benefit from fresh regional planning and new alliances is a matter for local study and conjecture, as well as private and federal advice.

For UAFs the regional mode is a natural one since their structure does not limit the outreach to single local governments or systems only. The 51 units in the present network are located in only 37 states and the District of Columbia; many UAFs already are in communication with multiple state health departments. In the regionalization prototypes presented in this chapter, UAFs had major involvement in single state (Ohio) and multi-state (New England, Boston) genetic projects. By virtue of their academic orientation, familiarity with new technology, and capacity for objective discussion and planning, UAFs are ideally suited to serve as catalysts and partners for the development of regional programs.

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II. A CONSIDERATION OF LINKAGES

Linkages Among UAFs and State and Local Title V Programs

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AAUAP MCH Consortium Project

The Foundations for Alliance

The bases for linkages between UAF programs and state Title V programs that are explored here reflect a wide range of areas of concern and cooperation on national issues and in the needs and priorities across the country. Table 1 summarizes information on alliances between a representative group of UAF programs and Title V programs, particularly state MCH and CC programs, in twenty-nine states and the District of Columbia. In some cases, these reports reflect long-established relationships; in others, the parties involved have only recently begun to explore and to initiate cooperative working arrangements. The linkages may be formal contracts or verbal agreements. They may be of short duration or may endure over a number of years. They may be periodic in nature, such as presentation of lectures within a university course, or may be more extended, as in instances where there is joint staffing or operation of specialty clinics. Consistently, at the core of each arrangement, there is the recognition of mutual benefit for the participating parties.

Just as the benefits are seen as reciprocal, so is the responsibility for initiating and sustaining the agreements. State Maternal and Child Health and Crippled Children's Services programs constitute the major component of the Title V program. Other Special Projects of Regional and National Significance, which include a number of the UAF network members, are established for support of the broad maternal and child health goals, including those of the state programs. Each has something to offer to the other and, indeed, dynamic relationships do not occur unilaterally.

Table 1. UAF and MCH/CC Linkages 1984
Degree of Involvement

- 1 - Periodic
- 2 - Moderate
- 3 - Substantial
- * - New or Emerging

Center for Developmental & Learning Disorders
Birmingham, AL

Division of Developmental Disabilities & Clinical Genetics
Irvine, CA

University Affiliated Facility
Mental Retardation Program
Los Angeles, CA

University Affiliated Training Program, Children's Hospital of LA
Los Angeles, CA

Georgetown University Child Development Center
Washington, DC

Mallman Center for Child Development
Miami, FL

Riley Child Development Program
Indianapolis, IN

Training							
Shared Trainees	1			*	1		1
Workshops/Seminars	3	1	1	2	1	2	3
Inservice Provided through Outreach	3	2		2	1	2	2
Extended Inservice at UAF Site	3			2			
Courses/Lectures	3	2	1	3	2	3	2
Trainee Practicum or Workstudy	1			1	1	3	*
Trainee Experiences in Clinics or Services	3	2	3	1	1	3	2
Service							
Operating MCH or CC Specialty Clinics or Services in UAF	3	3		*		3	
UAF Staffing of MCH- or CC- Operated Clinics	2	3	2	1			3
Follow-Up	3	3		3	2	3	
Outreach to Provide Service to MCH or CC Clients	3	3	3	2	2	2	
Service Provided on Contractual Basis	3	3		2		3	
Providing Service on Referral or Cross-Referral Basis	3	3	3	3	1	3	3
Advisory							
Special Topic Advisory Committee, e.g., Genetics, Developing Standards		3		2	3	3	3
Title V State Administering Agency		3			3	3	
Crippled Children's Program	2	3	*	3		3	1
MCH Program	1	3	*	2	3	3	1
SPRANS Program		3		2	3	3	1
Title V Personnel on UAF Advisory Committee	1						*
Interagency Committee for Communication or Liaison	2	3	1	3	2	3	
Staff							
Employment of UAF Trainees in Title V Programs	1		1	2	1	2	2
Shared or Jointly Funded Staff	1			3		2	2
Professional Consultation	2		2	3	1	3	2
Planning							
Developing Processes or Systems, e.g., Case Management, Needs Assessment	1	3	3	3		3	1
Regional Collaborative Projects	1				2	3	2
Research							
Utilizing Health Records		3	3	1	1		1
Shared Research			*	*		3	3

Division of Developmental Disabilities
Iowa City, IA

Kansas University Affiliated
Facility-Kansas City
Kansas City, KS

John F. Kennedy Institute for
Handicapped Children
Baltimore, MD

Developmental Evaluation Clinic
Boston, MA

Ennice Kennedy Shriver Center for
Mental Retardation
Waltham, MA

University Affiliated Facility
for Developmental Disabilities
Kansas City, MO

Meyer Children's Rehabilitation Institute
Omaha, NE

University Affiliated Facility
Rose F. Kennedy Center
Bronx, NY

Division for Disorders of
Development & Learning
Chapel Hill, NC

University Affiliated Cincinnati
Center for Developmental Disorders
Cincinnati, OH

The Nisonger Center
Columbus, OH

Child Development &
Rehabilitation Center
Portland, OR

Child Development Center
Memphis, TN

University Affiliated Center
Dallas, TX

Child Development & Mental
Retardation Center
Seattle, WA

Waisman Center for Mental
Retardation & Human Development
Madison, WI

	1	1		3	2			2			3	2							
3	2	2	2	3		1	2	3	3	2	3	3	3	3	3	3	2		
1	2	2		2	2	3	2	3	2	2	3	2	2	3	*		2		
	1	1		3				3		2	2	2	2	3	3	2			
2	2	1		3	3		2	3	2	2	1	3	3	1		1			
1	1	*			2					3									
1	3	3	3	1	3	2	2	2	2	3	3	3	3	3	1				
	1	3	3			2	3		3					3		3	3		
2	3	1	3	2		3			*	1	3	3	3	3					
3	3	1	1	2		2	3	3	3	2	3	3	3	3		3	1		
1	3	3	3	3	3		3	3	3		3	*	1	3	3				
1	2	3	2		1	3		3	3		3			3		3			
3	3	3	2	2		2	2	1	3	3				3	1	3	2		
1	2	2	3	3	1	3	2	2	3	3				3	2	2	3		
	1		3	*		2		2	3	2					2		2		
1	1	1	3	2	1			1	2					1	2	3	2		
1	1	1	2	1				1	2					3	3	3	3	1	
2			2	*					3	1				3	2	3			
	1	3	*	*		1			3							2	2		
	3	2			1		2	3	3	3	3	3	3	3	3	2			
	1	2	3	3			2	2	2	2	2	2	3	3		2			
1	2	2	1		1	2		2	2				3	2		2	1		
2	2	2	3	2	2		3	3	2	3			3			3	1		
	2	2	2	*	1	3	*	1	3	2	3	3	3	3	3	3			
2	2	*	3	2		2	*		2	1	3	2	2	*	*	*	2		
1	2		3	3					1		3		3			2			
1	1	3		*	1	2		1	2	2	2	3	*			1	2		

Collecting the Linkages Information

The UAF network was the vehicle for organizing the collection of information on a national basis. Reporting of the information was voluntary; however, joint reporting (between UAFs and state programs) was encouraged. As the conduit for the information, UAF personnel were asked to develop the information in concert with the State MCH and CC personnel or, alternatively, to achieve consensus with the appropriate state directors or their designees. It should be noted, too, that although this report refers frequently to state programs for MCH and CC, the organization of those programs varies in some areas. Thus, some UAF-CC linkages involve citywide CC programs, as in New York City, or regional programs, as in California. Except where specifically denoted as "the Ohio State MCH Program" or "the Maryland Crippled Children's Services," for example, the state program terminology is intended to refer to these varied structures as well.

Trends in the Linkages

As depicted in Table 1, the linkages that have been reported are summarized quantitatively. Without sophisticated statistical analyses, these data are more appropriately to be regarded as indicators than as absolute measures. In an attempt to capture the quality and, frequently, the unique character of the linkages, the following discussion focuses on selected vignettes of their content and their evolution. The discussion is organized according to the major components of collaboration (training, service, advisory functions, shared staff, technical assistance, planning and research) and will give special attention to new or emerging partnerships that have taken place in 1984.

Training

Diagnostic and treatment clinics have long been a staple of the state Title V programs' service delivery system. A number of UAFs are known to be

operating or jointly staffing such clinics. It has been known, too, that the state clinics are often utilized as sites for UAF training, although little has been documented about the practical considerations or outcomes of these arrangements. In an effort to explore the issue further, a study was conducted by the Core Curriculum Task Force of the AAUAP's Training Director's Council.

The first phase of the study was a survey of UAF Training Directors to determine the extent of usage of MCH/CC clinics for training, the types of clinics utilized, and the types of experiences afforded trainees within the clinics. Of the 34 Training Directors who responded, half reported that state funded clinics are used for training. The number of clinics used varies from 11 clinics used by one UAF to the use of one clinic by six UAFs.

Ratings of the effectiveness of the clinic sites in meeting UAF training needs were given by the respondents on a scale from 1 (Poor) to 10 (Excellent). Responses were as follows:

Rating of	4	by	1 UAF
	5		1 "
	6		1 "
	7		3 UAFs
	8		6 "
	9		2 "
	10		3 "

N=17

No UAF rated the effectiveness at below a "4" on the scale. Types of clinics reported as training sites include genetics outreach, nutrition, developmental or child development, physical rehabilitation (neuromuscular), communicative disorders, cerebral palsy, orthopedics, hemophilia, myelodysplasia, language disorders, craniofacial, cardiology, prenatal diagnosis and counseling, metabolic, PKU, educationally related therapy, newborn follow-up, spina bifida, comprehensive health care clinics, audiology and vision function.

Clinical training experiences were most often reported in the areas of

observation and participation in interdisciplinary team assessment, provision of service, and follow-up. Among typical training experiences reported were descriptions of genetic counseling interns acting as case managers coordinating services along with providing genetics counseling for the client or family, pediatric trainees functioning as case coordinator for the myelomeningocele clinic, provision of genetic consultation to other clinical teams and staff, and social work trainees organizing a Parents' Self-Help Group.

In the second phase of the study, the MCH and CC clinics identified as UAF training sites were contacted requesting their responses regarding the benefits and/or problems resulting from involvement in the UAF training program, arrangements for trainee supervision, whether or not the clinic personnel would again provide a training site, and their opinions on whether or not there is anything the UAF could do to improve and/or facilitate the training program arrangement. Of the 58 clinics contacted, 34 (59%) responded. A summary of responses on benefits and problems follows:

	<u>Yes</u>	<u>No</u>
Benefits for your Program?	28	6
Problems for your Program?	12	22

In describing benefits received, clinic personnel repeatedly noted that more service and more comprehensive service was provided. Eleven of 26 comments on benefits referred to increased service capacity. Other benefits mentioned were:

- o "new clients with different disorders" were seen,
- o development of additional procedures (follow-up protocols),
- o the presence of students was a stimulating experience for other staff,
- o a greater knowledge base of MR/DD clients,
- o improvements for staff in areas such as parent conferences, and
- o the inclusion of additional disciplines in the clinic teams.

Of twelve comments on "problems," seven noted space as a problem. Increased time demands on staff was mentioned by two clinics and one emphasized that provision of such a program "needs continued expansion, research, attendance at national and international meetings, etc., requiring increasing funding -- not decreasing."

Twenty-six clinics responded negatively to the question, "Is there anything the UAF could do to improve and/or facilitate the training program?" Eight responded affirmatively. Suggestions for improvement included increased predictability of student involvement, more written feedback to physicians and nurses, fewer numbers of students, more students, better orientation to medical students, orientation to parents about the importance of training, additional funding, and more regular involvement of UAF staff. Those that did not feel the UAF could improve the arrangements were either entirely satisfied with the current arrangements or acknowledged that present funding limited the extent to which changes could be made.

In addition to the use of clinic sites, other modes of delivery for training have resulted from UAF-MCH/CC relationships as well. Perhaps the most extensively utilized of these is the inservice workshop or seminar. Almost without exception, the linkages reported in Table 1 reflect cooperative activity in this category. The topics of the workshops/seminars have ranged from adaptive seating to management of high risk infants, from augmentative communication systems to dealing with intellectually limited mothers.

In the District of Columbia, for example, the Georgetown University Child Development Center worked collaboratively with the DC CCS to plan and present a workshop, "New Perspectives in Diagnosis and Management of High Risk Infants and Young Children." More than 300 health care professionals attended the workshop which was CME accredited. A similar workshop was planned between GUCDC and representatives of the Virginia Bureau of Maternal and Child Health

focusing on the high risk infant. Again, more than 300 health care professionals attended. In each case, there was broad representation from the state Title V-CCS programs.

On a monthly and bi-monthly basis, inservice training is provided by the GUCDC to assist community clinics in integrating prevention programs into the routine of normal health care delivery. Statewide continuing education on prevention for rural health professionals and teens and young adults is provided in Ohio through a collaborative organization involving the University Affiliated Cincinnati Center for Developmental Disabilities (UACDD) and Nisonger Center UAFs, Area Health Education Centers, and Ohio Cooperative Extension Service.

In Indiana, the Division of Maternal and Child Health has requested participation of the Riley Child Development Program UAF staff in a major workshop for state MCH ambulatory preventative care project staff. The theme of the workshop is early identification of developmental disabilities. In Iowa, a workshop to share program knowledge and to explore "current issues" in delivery of health care to disabled children was held by the Iowa MCH and CCS programs and the Division of Developmental Disabilities UAF involving approximately twelve staff from each program. On-site training was the program involving three nutritionists from the Kentucky and Tennessee MCH/CC programs, each of whom spent a month in the clinical training programs of the Child Development Center UAF in Memphis.

Cooperation involving the Massachusetts Departments of Social Services, Mental Health, and Public Health and the UAFs (the Children's Hospital Developmental Evaluation Clinic and Eunice Kennedy Shriver Center for Mental Retardation) resulted in a workshop focusing on issues and needs of limited parents. Intellectually limited mothers were the focus of a conference co-sponsored by the Rose F. Kennedy Center UAF, the New York City Department of

Health Bureau for Handicapped Children and the New York State Bureau of MCH, Special Services Children's Unit. In the northwest, a consortium of UAFs and state programs planned and conducted a workshop on "Early Intervention: The State of the Art" as part of the agenda of the Northwest Developmental Pediatric Group. The planning committee involved representatives of the Oregon MCH and CC programs, the Idaho CCS program (chairperson of the region's MCH/CC Directors group) and UAFs in Seattle, Washington, and in Portland and Eugene, Oregon.

As mentioned, the incorporation of single lectures within university courses is not infrequently used as a way to reach various disciplines within the health professions. At the UAF of South Carolina-Rock Hill, the UAF staff present lectures in courses in area nurse training programs. Full courses are developed as well for Title V personnel, such as the 15-week graduate level course, "Developmental Disabilities: An Interdisciplinary Approach," offered by the Georgetown University Child Development Center to professionals from D.C. MCH clinics. Approximately ten Title V CCS personnel participate in the course each semester.

The Kansas UAF-Kansas City worked jointly with the Division of Health of the State Department of Health and Environment, the Division of Continuing Education, and the Department of Pediatrics of the Kansas University Medical Center to develop and offer an outreach course in developmental disabilities for physicians and allied health professionals in the state of Kansas. The course was offered in the fall of 1983 and again in the spring of 1984. As part of a three-year project funded by the Public Health Service, the Kansas Department of Health and Environment has contracted with the Kansas UAF-Kansas City for assistance in the coordination and delivery of training and the evaluation of educational activities for the Kansas Genetic Service Project.

For long-term training, the Children's Medical Services of Florida has

invested in interdisciplinary training with funding of a program at the Mailman Center UAF, to complement the existing federally funded training program. State funding has been made available to extend the scope of the interdisciplinary training program of the Riley Child Development Program UAF in Indianapolis as well. State Maternal and Child Health funds are available for nutrition fellows who rotate through the Riley program.

Service

As mentioned previously, a number of service agreements are in effect for the operation of state programs' clinics through UAFs. Examples include:

The Massachusetts Infant Follow-up and PKU clinics operated by the Developmental Evaluation Clinic UAF at Boston Children's Hospital

Waisman Center UAF operates the Wisconsin State Metabolic Clinic which coordinates a newborn screening program, provides screening for Tay Sachs disease, and provides care for maternal PKU

KUAF-Kansas City operates the NICU Follow-up Clinic for the Kansas Department of Health

Some interactions defy simple categorization, involving a variety of components. In each, however, service delivery is a common basis for cooperation. One example is the Early Childhood Intervention (ECI) Program in Texas, a statewide interagency program to provide screening, assessment, referral, and infant stimulation services for children from birth to three years. The University Affiliated Center in Dallas has played several roles in the development of the ECI program, from planning (representation on the Texas Advisory Council on ECI) through implementation (providing screening, assessment, and referral services as well as consultation to service programs statewide). The UAC also contracted to develop educational modules for professionals working with developmentally handicapped infants and children and their families.

MCH Services of the Oklahoma State Department of Health (OSDH) and the University Affiliated Center (UAC) in Dallas have jointly planned an inter-

agency approach, at both state and community levels, for early intervention services that are integrated across agencies. The UAC will facilitate the development of interagency councils at the state level and in two pilot communities during the first year of the two-year project and will serve as a preceptor for a Coordinator for Early Intervention Services, a new position within OSDH which will provide continuity for continued development of state services beyond the grant period.

Within Massachusetts, the Division of Family Health Services (DFHS) of the Massachusetts Department of Health has joined with the Developmental Evaluation Clinic UAF of the Children's Hospital and the Harvard School of Public Health to conduct a policy development and demonstration project to look at the most appropriate content, organization, and financing of services for families with handicapped and chronically ill children. While Project SERVE addresses an area of current national discussion and debate and is federally supported, the project will directly assist the DFHS to define its present and future role for the delivery of services for handicapped and chronically ill children.

Staff

A corollary to clinic operation is found in staffing patterns that evolve through agreements between the state programs and UAFs, as in California, where staffing of CCS Medical Treatment Units is shared with UAFs at the University of California-Irvine, Children's Hospital of Los Angeles and, effective in the fall of 1984, the University Affiliated Facility Mental Retardation Program (Los Angeles). The Medical Director of the State Crippled Children's Services of Alabama also serves as Director of the Division of Medicine of the Center for Developmental and Learning Disorders UAF in Birmingham. In Rochester, New York, the County Health Department pays the salary of a social worker who works in the Birth Defects Center at the UAPDD.

Provision of service and shared or jointly funded staff are combined in a number of other cases, as in...

Washington State, where the MCH Program contracts with the Child Development and Mental Retardation Center (CDMRC) UAF to provide one nurse whose time is split equally between the High Risk Follow-Up clinic and continuing education responsibilities;

Florida, where offices and clinics of the Children's Medical Service are located at the Mailman Center UAF, which allows for an ongoing partnership that includes shared staffing and operation of clinics. Current state-sponsored programs include the Child Protection Team, the High-Risk Follow-up Program, the Developmental Evaluation (High-Risk) Model Program, Genetic Services Program and the Child Development Clinic;

Iowa, where pediatric faculty have served as examiners within the Specialized Child Health Services regional evaluation and planning clinics;

Nebraska, where the Meyer Children's Rehabilitation Institute UAF has provided services to the State Services for Crippled Children's Cerebral Palsy, Myelodysplasia, and Educationally Related Services clinics. The agency has also contracted with MCRI to provide the SSI-Disabled Children's Program statewide. Case management in the state has been enhanced by an MCRI-developed computer program for follow-along; and

Tennessee, where the Inborn Errors Clinic is located in the Child Development Center UAF and dental staffing is provided by CDC for the Hemophilia Clinic.

All dental plans submitted to the Kansas Crippled Children's Program for authorization are reviewed by the Director of the Pedodontics Division of the Kansas UAF-Kansas City. In Ohio, the Nisonger Center has cooperated with Children's Hospital in establishment of the NICU Follow-up Clinic at Nisonger and for collaborative inservice training in Level II nurseries. Follow-up evaluation for the Cerebral Palsy Clinic of the Wisconsin SSI-Disabled Children's Program is provided by the Waisman Center.

Through an agreement between the Utah State MCH Program and the Exceptional Child Center UAF at Utah State University, approximately 100 children will be evaluated for whom treatment plans will be developed jointly by the State MCH and UAF programs. The UAF at the University of Southern

Mississippi is working with the local district health department and the regional MR/DD agency to coordinate services, with shared staff, for the assessment of handicapped children. Beginning in the fall of 1984, UAP trainees will be involved in the program as well.

The New York State MCH Program has contracted with the University Affiliated Program for Developmental Disabilities (UAPDD) in Rochester and the Regional Perinatal Center to do Infant Health Assessment Checks. UAPDD has received one of four state MCH grants to demonstrate coordination of care for chronically ill and physically handicapped children, which creates an interdisciplinary team to develop IHPs for children seen in subspecialty clinics of Strong Memorial Hospital and through staff shared with the county MCH program, to assure referrals and information provision through outreach from Community Health Nurses. The project also contains a continuing education component.

Advisory Relationships

The nature of advisory relationships has also been explored intensively for this review. As can be seen in Table 1, most of the advisory categories are self-explanatory, e.g., the meaning of "Title V personnel on the UAF Advisory Committee" needs little interpretation. An interesting overall pattern in the data is that the advisory role seems skewed toward UAF personnel advising state programs rather than the other way around. Not shown in the table is an instance in which the state program advisory membership is rotated, e.g., whereas last year the Child Development Center UAF in Memphis was represented on a Title V advisory group, that position has now devolved to another program in the state.

Also not reflected on the chart is membership of the Kentucky MCH Program Director on the Human Development Program UAF Advisory Board. The HDP, which is not a DMCH-funded program, is representative of several programs which are referenced throughout this report and for whom the Title V relationships are

relatively new in all cases. Those programs include, in addition to HOP, the UAF of South Carolina, the University Affiliated Program of Mississippi, the University Affiliated Program for Developmental Disabilities (Rochester, New York) and the Exceptional Child Center UAF (Logan, Utah). Certainly, these programs have not had the historical MCH linkage of other programs but, regardless of the extent to which the programs cooperate, they affirm that richly productive, mutually beneficial agreements are not limited to traditional categorization by funding source.

Advisory functions around special issues are a frequent vehicle for pairing the needs and skills of UAFs and state MCH and CC programs. Some of the occasions for this include:

Partnership of the County CC program and the University Affiliated Training Program, Children's Hospital of Los Angeles, for standards for pediatric home health care;

Development of orthopedic protocol for CCS clinics (Meyer Children's Rehabilitation Institute, Omaha);

Development of new State Health Standards for Children with the Tennessee MCH Program and representation on the state committees for genetics and perinatal services and the Governor's Task Force on Healthy Children (Child Development Center UAF, Memphis);

Participation by Waisman Center UAF staff in Maternal PKU Planning (to formulate standards and policies for statewide maternal PKU program), in addressing the problem of a comprehensive state registry for children with PKU (PKU Follow-Up Program Work Group), in the Genetics Planning Committee to address strategies to enhance the statewide genetics network, and through the Statewide Prevention Task Force, established by the Waisman Center, the Wisconsin State DD Council, and Title V agencies to develop statewide prevention policy recommendations;

The Massachusetts Advisory Committee on Genetics Services includes the PKU Clinic Director (Developmental Evaluation Clinic UAF);

The University Affiliated Cincinnati Center on Developmental Disabilities UAF has membership on Ohio's MCH/BCCS Committees on Myelomeningocele Standards, Cerebral Palsy Standards, and DD Nursing Care Standards; and

The Mailman Center UAF is represented on the Florida Infant Screening and Genetics Advisory Councils.

A Final Note

The foregoing has been an attempt to provide an overview by example of the types and extent of linkages/agreements/working arrangements between the members of the UAF network and state Title V programs. Selections to highlight the discussion have been chosen for their power to illustrate the issues and areas of interactions. With the great amount of information available, the reader should not infer that this tells the entire story for any program or collaboration. In fact, these descriptions are deliberately abbreviated, and in each case, a great deal of material could be devoted to completing the details. That is done elsewhere and for other purposes. Our purpose is to highlight and enlighten, to stimulate and encourage, and to pique interest among those who could be participating in such arrangements and are not. The bottom line is that these relationships lead to improved services to developmentally handicapped and chronically ill persons regardless of whether the focus is on such pedestrian topics as how to initiate and develop staff sharing agreements, cooperation for preservice, inservice, or continuing education, or the numerous other areas explored herein.

The final paper in this book speaks directly to this last point. MacQueen, in describing one system, stresses the integration of the services delivered rather than the integration of administration of services. His paper, for our purposes, might be said to be "where it's at" for the future of service delivery.

The study referenced on page 104 is an unpublished report of the results of a survey conducted by Calvin Knobloch, Training Director, Division for Disorders of Development and Learning, The University of North Carolina-Chapel Hill, and Mary Ann Smith, Training Director, Child Development Center, The University of Tennessee Center for Health Sciences.

THE INTEGRATION OF PUBLIC SERVICES FOR HANDICAPPED CHILDREN:
MYTH OR REALITY

John C. MacQueen, M.D.
Iowa Specialized Child Health Services

Adapted from a presentation to the "Health Care for the Handicapped Child: Sharing Solutions to Common Problems" Seminar, March 28, 1984, Memphis, Tennessee

In 1934, there were many children in this nation who had been physically crippled by such health problems as paralytic poliomyelitis, cerebral palsy, and congenital skeletal anomalies. Few of those children had access to the rehabilitation services they needed, however, because the private medical system was not prepared to provide those, and there was no public medical system involved with providing the services. Crippled children who lived in rural areas had essentially no access to rehabilitation services. There was great concern on the part of those responsible for providing health care for children and on the part of the general public that the then new methods to diagnose, treat, and rehabilitate handicapped children were not available to the crippled children of the nation. At that time, the medical specialty of orthopedic surgery was in its infancy and the American Academy of Orthopedic Surgeons listed only 471 members. The concern of the public resulted in the creation of what was then a new and revolutionary idea--that each state should have a public health program that was specifically concerned with crippled children. The idea was succinctly stated in Section V of the legislation:

"for the purpose of enabling each state to extend and improve (especially in rural areas and areas suffering from severe economic distress) as far as practical under the conditions in such state, services for locating crippled children, and providing medical, surgical, corrective, and other services and care and facilities for diagnosis, hospitalization, aftercare, for children who are crippled and who are suffering from conditions which lead to crippling...."

separate program as though the other programs for handicapped children did not exist. Each was organized to function parallel to other programs, but not to be coordinated with other programs. Each of the programs had its own constituency. Each program was designed to have its own legislative mandate, its own administration, its own regulations, its own staff, its own eligibility, and, as a result, its own turf. The result is an unbelievable fragmentation of services for handicapped children. In some instances, the fragmentation has resulted in a duplication of services and thus in a duplication of cost. In some, it has resulted in gaps of services. It has caused programs to compete with other programs for the same population of children. And, in some instances, a single child has received services from multiple agencies for the same problem. But, most importantly, it caused confusion on the part of parents and so resulted in underutilization of services.

During recent years, the problem has been described in many studies and has been recognized by members of the Congress who have been involved with the oversight committees responsible for these programs. The state governors became particularly concerned about the problem in the late 1970s when they came to realize that not only was a significant part of their state budget being used as matching funds for federal programs that were not always in keeping with the state's priorities, but the states could not change the program as needed to develop coordinated services, and so they were required to accept the fragmented services created by the federal legislation.

It is to be remembered that early in his Administration, President Reagan requested the approval of state governors for his policy of returning the control of these programs to the states and they overwhelmingly supported the idea. Professionals working in these programs have many valid reasons to support the programs. I would urge, however, that loyalty need not cause us to support the manner in which the programs are designed when that results in

fragmentation. It has been said that the only people who aren't aware of the fragmentation are those of us who provide the services--those of us who create the fragmentation.

Until the passage of the block grant legislation, there seemed to be no solution to the problem of fragmentation caused by multiple federal categorical programs. However, with the enactment of this legislation, the state legislators and agency officials are now responsible for appropriating these funds and so for determining how the money can be spent. It is now possible for them to change programs so that they will be not only more effective but more economically efficient. It is a matter of record that the state CC programs are in a period of transition when the state legislatures are, for better or worse, making changes in the programs to reduce the fragmentation of services--to coordinate the programs that provide services for handicapped children. Let me describe what is probably the major example of a lack of coordination in the provision of services for handicapped children.

Soon after the state crippled children's programs were created in 1934, it became evident that the services needed by crippled children must be provided by medical specialists and by a professional team so the services could not be provided by a local health department or by public health nurses, in contrast to the services provided by many MCH programs. For that reason, the state crippled children's programs were developed to provide services in a variety of settings that included specialty clinics and/or the offices of the professionals involved with providing services. These arrangements were satisfactory for providing diagnostic, consultation, and planning services for handicapped children but they were not satisfactory for providing day-by-day rehabilitation services. This problem is shown by the following. In the past, if a generally trained physician referred a child to a crippled children's service, the child was seen by a specialist, a diagnosis was made

and a plan for rehabilitation services was proposed. That plan might have included speech and hearing, occupational therapy, physical therapy, psychological services, etc. However, all too often, the child could not obtain those services in his or her community. Such community services just didn't exist in the great majority of American communities. Where they existed, they were most often supported by voluntary health organizations such as United Cerebral Palsy, and these units varied a great deal in their comprehensiveness and in the quality of services they provided. In particular, it is noted that, in the past, the public school system was not involved with providing services for these children. The stark truth of the matter is that, because the CC model does not provide community based rehabilitation services, the great majority of handicapped children did not have access to complete services. This situation persisted until the passage of P.L. 94-142 required the public school system to develop and conduct community based rehabilitation services for these children.

It would be assumed that, in 1975, those responsible for creating a new community based system of rehabilitation services to correct the previously described situation would have written legislation to coordinate the new community based system with the long established CC public program that provides diagnostic and treatment services for the same population of children. But, no, in keeping with the policy or habit of creating categorical programs as though no other program existed, P.L. 94-142 was written to make no reference to the CC program, nor does it mention the physician's responsibility in providing services for handicapped children. The word "physician" does not appear in the legislation. It does make reference to the health team of which the nurse is a member. Thus, when this very important legislation was designed, when there was a possibility for the first time to put together a comprehensive public system for providing services for handi-

capped children, those responsible goofed. The rules of the game of categorical programs overruled good judgment. As a result, we have on one hand a program that is able to provide identification, diagnostic, treatment, and planning services for handicapped children (the CC program) and, on the other hand, we have a community based system designed to provide rehabilitation services for handicapped children. And, because the legislation does not require or suggest cooperation, I can assure you that, throughout this nation, little effort has been made to coordinate the services provided handicapped children by these two programs.

This is only one example of the lack of coordination that exists between all categorical problems. It is a nationwide problem. Now we must turn our attention to how this problem can be solved.

Because the categorical programs have been created by federal-state legislation, it is easy to assume that the solution to the problem would be to change the legislation. However, if we try to solve the problem by going the legislative route, it will be necessary to be involved with the political process at the federal and state levels to rewrite a great deal of legislation. And, I would suggest that, even if we are successful, that would take a very long time.

In lieu of this, I would suggest a very different solution. It is a solution that operates within the framework of the current legislation. It is a solution that creates a state-community based system to provide services for handicapped children.

It is my belief that it is our purpose to coordinate and integrate the services provided each handicapped child. It is not our purpose to coordinate and integrate the administration of the programs that provide the services. These are very different purposes. Because our purpose is to coordinate and integrate the services for the individual child, we must be involved with

developing arrangements between the public agencies that provide services for the individual child in the community.

To describe philosophically what arrangements should be made in the community to coordinate services for each child could be very time consuming so I have elected to describe one state's experience--not as a model but as an example of what arrangements can be made. Hopefully, it will give you some ideas that you can apply or adapt in solving problems in your own state and in your own community.

In 1976, the Iowa Crippled Children's Service became convinced that its mobile field clinic program was an excellent program for making subspecialty level diagnostic, planning services, and followup services available to children of that rural state for those children who were to receive their treatment services in a medical center. However, the mobile field clinic program was of little value in providing services for children with developmental, behavioral, and educational problems who were to receive their treatment services from local professionals in the community. We summarized what we believed to be the problem and a solution to the problem in a document entitled "The Provision of Contemporary Child Health Services Through Community Child Centers." That document proposed that in order to provide contemporary services, it would be necessary to develop regional child centers. We created centers in two rural Iowa communities. In short order, we learned a great deal about the problems of organizing such centers but we also became more convinced that there was a great need for a community based service system.

In 1978, the federal Office of Maternal and Child Health and the federal Office of Education developed a program to support projects that would demonstrate methods to coordinate health and educational services in the community. We applied for and received one of the competitive grants. Our experience

rics recently visited the Ottumwa Center and, in the near future, the Committee will be making a positive report to the Executive Board of the AAP describing the Center and indicating their belief that the Regional Center program is an interesting possible solution to the problem of providing services for handicapped children.

In addition, there have been related legislative activities. Two years ago, a request was made to the legislative committee responsible for the state CC program to charge those of us who are involved with the provision of services for handicapped children with conducting a study to evaluate what we are doing and how we could do it better. As recorded in the Journal of the House of the Iowa Legislature, the charge read:

The state department of health, Iowa specialized child health services, the university of Iowa hospitals and clinics, and the department of social services shall jointly study and develop a plan for the integration and coordination of maternal and child health programs, including but not limited to prenatal clinics; obstetric clinics; maternal health centers; child health centers; well-child clinics; the women, infants, and children nutrition program; the maternity and infant care project; the children and youth project; dental clinics; specialized child health clinics; related medical assistance programs, including the early and periodic screening, diagnosis, and treatment program, and medical assistance reimbursements for maternal and child health services; and county maternal and child health programs. The plan shall provide, if possible, for locating the clinics at the same sites and for the sharing of administrative expenses. The plan and proposed implementation schedule shall be developed and submitted to the joint human resources appropriations subcommittee by January 31, 1983.

The involved departments of state government subsequently conducted the study, and it proved quite valuable in that it caused us to better understand what each other was doing and to begin to make changes to decrease the fragmentation in the services we were providing.

One of the recommendations of the study was that we should develop demonstration projects in two communities--Sioux City, which is a relatively large community, and Ottumwa, which is a smaller community--to explore and demonstrate methods of interagency coordination. This was done last year. It was

a very successful project and the legislative oversight committee has charged us to continue that activity and to expand the program to include two more communities during the year.

This brief report about our legislative activities is included to emphasize that in establishing a new system, it cannot be limited to conducting a few demonstration projects. It must involve all those units of government that are responsible for providing services for handicapped children. It must address the difficult problem of changing attitudes. It must include the private medical community. It must be institutionalized so that all parties come to expect that it will function as part of the total system. It must alert state legislators that it is possible to conduct more efficient and more effective programs to provide services for handicapped children.

Because the concept of categorical programs is so much a part of the accepted organization of state government and, because fragmented services are too often accepted as the norm, a revolutionary change will be required to achieve the goal of integrating services for handicapped children. But, lest the idea of a revolutionary change be frightening or disheartening, I would remind you that the creation of the state crippled children's program in 1934 was considered revolutionary.

So, in response to the question--"The Integration of Public Services for Handicapped Children: Myth or Reality?"--I believe it can be a reality but we cannot wait for the process to start. We cannot wait for the governor or for the commissioners to start the process. It must start with people who are informed about and dedicated to the provision of services for handicapped children. It must start with you.

The motivating force in the development of the state crippled children's programs was frequently a well-known orthopedic surgeon. I can personally testify to this because I well remember that Dr. Francisco, an early orthopedic surgeon from Kansas City, brought a state crippled children's clinic to the small Kansas town where I lived as a child and cared for my older sister who had a minor orthopedic problem. It is difficult to describe to you the aura of importance and the sense of gratitude that my family and my town felt for Dr. Francisco and his clinic. Because of such men as Dr. Francisco, many state crippled children's programs were developed to provide only orthopedic services. In fact, as recently as a few years ago, several state crippled children's programs continued to limit their services to children with orthopedic problems. However, within a few years after they developed their CC programs, the majority of states recognized that a child's ability to function could be limited by a chronic or recurring illness just as their ability to function could be limited by an orthopedic problem, and as a result many states soon expanded the definition of "a crippled child" to include chronic and recurring medical problems.

In more recent years, our society has recognized that there are a large number of problems, currently referred to as "the new morbidity," that can be handicapping and, as a result, many state CC programs have now expanded their programs to provide services for children with developmental, behavioral, and psychosocial problems. Most recently some CC programs have elected to provide home-community services for children who have complex health problems that require not only the traditional general support but high-tech care as well. An example of such children is the ventilator-dependent child. To provide services for this new generation of problems will require major changes in the state CC programs for they are not staffed, organized, or funded to provide such services. The point is that whereas in 1934 the state CC programs were

limited to providing services for children with orthopedic problems, the majority of state CC programs now provide services for children with a wide variety of health and health-related problems.

This is not to suggest that all services for handicapped children have been, or are now, provided by the state CC programs. In fact, our nation has seen fit to create a large number of categorical public programs to provide services for children with certain diseases and disorders and, in 1975, Congress passed the Education for All Handicapped Children Act that requires the public school system to provide a wide spectrum of services for handicapped children.

As noted, in the 1930s it was recognized that there were children crippled by orthopedic problems who needed special services, but it is now recognized that there are children with a wide spectrum of health and health-related problems who need special services. It becomes evident that the word "crippled" is no longer an appropriate name for these programs. Unfortunately, we have not decided on a new name for this population of children. Handicapped children, disabled children, children with chronic diseases and disorders, and children who need special services have all been recommended. I elect to use the term "handicapped children" for this paper.

Whatever the name given this population of children, the truth of the matter is that, at this time, in the 1980s, many American children--far too many American children--who have chronic health and health-related problems do not have access to needed services. To assure that these children have access to contemporary services remains the unfulfilled responsibility of our society.

During the past 50 years, many physicians, nurses, occupational therapists, physical therapists, speech and hearing personnel, psychologists, and

social workers have provided services for handicapped children and, from that experience, a body of knowledge has been developed about the modern technical services needed by handicapped children with different handicapping diseases and disorders. A consensus has also been developed that these services should be provided in a unique supportive fashion if they are to be effective. The following standards of care have been agreed upon:

- care must be family based,
- care must be personalized,
- each child must have an individual plan of care,
- coordinated interprofessional team care must be available because most handicapped children have more than one problem,
- one person will be responsible for assisting the family to carry out the program, and
- the cost of the program must not destroy the family's finances.

Thus, both the technical services and the support services required by handicapped children are very different from the services and the characteristics of the services required by children with acute illness. These differences are so great as to make it necessary to organize special systems to provide services for handicapped children.

Thus, in the 1960s, when this nation became involved with the idea that it could and therefore should create massive federal public programs to solve its social problems, Congress saw fit to create a number of new systems--new categorical programs--to provide services for unserved or underserved children with handicapping conditions. A recent study of the programs, as conducted by the United States Senate, identified some 60 such programs. Medicaid, EPSDT, SSI-DCP, the Genetics and Hemophilia Programs, and P.L. 94-142 are just a few of the major programs thus identified. Each of these programs was created for a good cause and each deserves our support. However, each was created as a

with the two regional centers had shown us that our public health nurses and other administrative staff did not have the experience needed in community development so we employed personnel with specialized training in community organization to organize a statewide system of regional councils and develop the service programs in each center. The organization is shown in Appendix 1. Each of these regional centers varies a great deal. Some are incorporated. Some are loosely defined. The services that are provided in the centers vary a great deal depending on the community's perceived need for services and on the interest and skills of the Center's personnel.

The core staff of the center is composed of a Pediatric Nurse Practitioner with special training in the care of handicapped children, a secretary, and a private physician who has received additional training in our UAF in the care of handicapped children and who is interested in conducting a special interest practice in providing services to handicapped children. The remainder of the staff in the center are provided by the Area Education Agency and by the Department of Human Services. Thus, the center does not duplicate personnel but serves as a meeting place where the representatives of the community agencies that provide services for handicapped children can meet to work together to evaluate a child's needs and to establish a plan of community services. A person is designated to be responsible for the services listed in the plan, and that person is also responsible for providing case management services. Thus, the center achieves the goal of coordinating the services for the individual child by coordinating the services that are provided by community agencies.

The program or activity conducted in a center that may be of greatest interest to you is the Integrated Evaluation and Planning Clinics. In these clinics, we see children who, for the greater part, have developmental, behavioral, and educational problems. The majority are referred by the school

system. The statistical information on types of problems seen in these clinics is as follows:

Behavioral	40.8%
Retardation & Developmental	13.8
Orthopedic	3.7
Speech & Language	6.0
ENT & Hearing	4.5
Cardiovascular	3.2
Respiratory	3.0
Other	14.3

These clinics have been very successful. They have the support of the public education/special education program, primarily because the clinics give the school direct contact with a physician who is interested in children with this type of problem, who can freely discuss with them the medical/health implications of their problems, and who functions as a member of the community team.

A few years ago, it would have been impossible to have created the centers because it would have been impossible to recruit the physicians needed to work in the clinics. During recent years, there has been an increase in the number of pediatricians entering practice in the state and an increase in the number of young pediatricians looking for a special interest practice. The regional clinics provide them with the support needed to conduct a special interest practice in developmental pediatrics.

Review of this program is provided to show that it is possible to create the community based arrangements needed to coordinate the services provided each handicapped child; however, you may question whether or not this describes a demonstration project. In fact, we now have state financial support for this system of clinics and the communities provide significant fiscal support for the regional centers. So, we've gone a long way toward institutionalizing the concept.

The Committee on Community Pediatrics of the American Academy of Pediat-

A PROPOSED SYSTEM TO PROVIDE CONTEMPORARY CHILD HEALTH SERVICES

